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A CONTRIBUTION TO THE STUDY OF MYOSITIS OSSIFICANS PROGRESSIVA.

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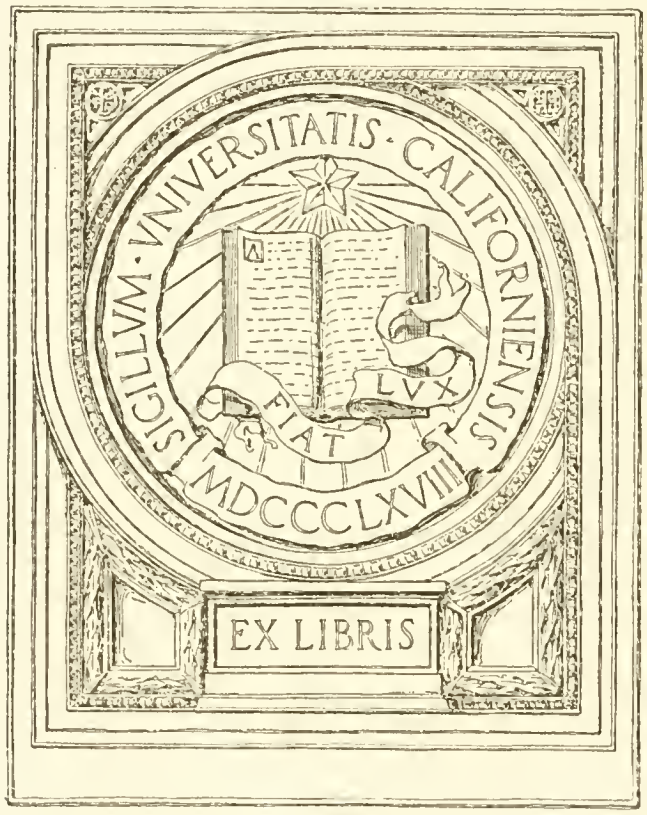

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A CONTRIBUTION TO THE STUDY OF MYOSITIS OSSIFICANS PROGRESSIVA

By Julius Rosenstirn, M.D.
of San Francisco
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(From Mt. Zion Hospital, San Francisco, Calif.)

With the first case of myositis ossificans progressiva described in medical literature over two hundred and twenty years ago, and for sixty years the course and pathology of this strange and rare malady well studied, its character and etiology are still unsettled and the subject of vivid controversy.

The fact that another resemblant disease, the myositis ossificans circumscripta, produces identical pathological anatomical tissue changes in isolated muscles has not helped to clear up the mystery of its etiology and certain features in the symptom picture. The myositis ossificans progressiva is characterized by such a sharply outlined group of symptoms that it is marked as distinctly etiologically separate from its near namesake and not sharing anything with it in common save a relationship in local products of the pathological process.

These characteristic features are:

1. The ossification of muscles without any apparent cause, traumatic or otherwise.

2. The manifestation of the disease as a congenital one or appearing early in life.

3. The progressive course of the malady unaffected by therapeutical efforts, embracing in some of the most advanced cases nearly the entire voluntary muscular system.

4. The association with symptoms of defective anatomical and physiological formation, mostly of fingers and toes but not infrequently also of stature, habitus and sex differentiation.

These are the earmarks of typical cases of myositis ossificans progressiva, and, strange to say, there are very few exceptions, if any, of genuine cases of this malady which do not show them.

It is true that since Helferich first drew attention to the presence of microdactylia in patients suffering from progressive ossifying myositis, quite a number of histories have been published that do not mention this symptom in cases undoubtedly belonging to this malady, but I have ascertained through personal correspondence with the authors of a number of such publications that the reason for this has been a lack of attention in examining for this symptom, mostly due to an oversight in the outdoor department and also in a few hospital cases.

There are, however, a few cases where the authors have especially mentioned the absence of microdactylia, or similar anatomical defects, and these are the observations which deserve special attention. Under the heading "remarks" of the collected case histories, the reader will remember this difference between *microdactylia not mentioned* and *microdactylia noted as absent.* The later definite statement should, in future, accompany all histories of such cases verified by the X-ray pictures of the hands and feet.

I have taken the trouble in compiling in chronological sequence all the accessible cases of myositis ossificans progressiva. It seemed to me to be an effort well worth while to assemble in brief all the known data of this rare and mysterious disease, as my studies of its literature disclosed many errors and omissions in the two or three fractional prior attempts.

The reason for undertaking this work was my own observation of a most interesting typical case, presenting among its symptoms the unique feature of several isolated and independent foci of ossification in the skin, a tissue hitherto believed to be exempt from this pathological change. It afforded unprecedented opportunity to observe and study the entire course of ossification from its first beginning to the completed bone-formation, a study which led me to advocate a change of view as to the primary factor of ossification in this disease.

In the arrangement of this paper I shall first give abstracts of all cases of myositis ossificans progressiva published to date, then present my own case and finally discuss the views of the pathology and etiology in the light of knowledge obtained by the study of the existing literature and of the findings of my patient.

ABSTRACTS OF CASES RECORDED IN LITERATURE

Case I.—Reported by Guy Patin in 1692. Female. Subsequent course of disease unknown. Writes to A. F. (?) referring to a previous letter sent him, wherein he also mentioned the case of a woman who finally became as hard as wood all over.

Case II.—Reported by John Freke, in 1743. Male, aged 14 years. First symptoms appeared at 11 years of age—swellings on back, muscles and vertebræ of back. Began with swellings three years ago, then growths spread over entire back from cervical vertebræ to os sacrum, and arising also from every rib, forming coral-like ramifications. Third year, from nape of neck to os sacrum and the lateral muscles of back; ossifications like coral branches, especially on back. No congenital abnormalities mentioned.

Case III.—Reported by Rev. Dean Copping and Robert, R. R., Lord Bishop of Corke, and Charles Smith, in 1744. Skeleton of man, aged 60 years. First symptoms appeared at (?) 18 years of age. Subsequent course of disease unknown. Entire

skeleton one mass. Could not open jaws, fed through gap in teeth. Vertebræ consolidated by lateral outgrowths with scapula. One bone, very thin, about four inches long, found in fleshy part of arm, *quite distinct and disengaged from any other bone in his body.* Ridges and reefs of bone through fleshy parts of both thighs like shoots of coral, eight to nine inches long. From back of calcaneum grow spurs of bone. No congenital abnormalities mentioned.

Case IV.—Reported by William Henry, in 1759. Male, aged 19 years. First symptoms appeared at 17 years of age—right arm, swellings and pains in wrist. Both arms, ankles, and legs up to knees attacked. Both arms from elbows to wrists just like a solid mass of bone; later pain, swellings and ossifications attacked ankles, creeping up fleshy part of calves to knees. Was treated with mercurials to salivation. (Observation covering two years.)

Case V.—Reported by Abernethy, in 1830. Male, aged 14 years. First symptoms and age at their appearance unknown. Various bony masses had formed at different times and disappeared again. Back greatly deformed by irregular hillocks of earthy matter over processi spinosi. Head immovably fixed backward and to one side. Exostosis on both ossa brachii, and tendinous margins of axillæ ossified so that arms were pinioned closely to his sides. Exostosis on pelvis between os sacrum and coccyx. No congenital abnormalities mentioned. Abernethy found urine contained less lime than normal. Two years later he gave patient phosphoric acid, 4.0 per diem in divided doses, with the result that normal quantities of phosphates of lime were excreted in the urine. When phosph. acid was discontinued deficiency of lime occurred again.

Case VI.—Reported by David L. Rogers, in 1833. Male, aged 13 years. First symptoms appeared when at the age of 12 years and 6 months—stiffness in arms and neck. Very rapid course, attacking muscles of back and chest. Superior portion of pectoralis major and sternocleido from sternum to middle portion ossified. A number of osseous swellings in back. Scapula fixed to ribs, studded with bony excrescences; all muscles connected with scapula—viz., trapezius, rhomboideus, subscapularis—more or less ossified; latissimus and longissimus dorsi formed large bony plates from their origin to angle of scapula. No congenital abnormalities mentioned.

Case VII.—Reported by Testelin, and Charles Dambesi, in 1839. Male, aged 26 years; mother suffered from rheumatism; patient fell on right thigh. First symptoms appeared at 18 years of age—pains and limping after trauma. Right thigh attacked. Five years later pains in all extremities, movements began to be difficult, particularly scapulo-humeral articulation. Continued to move around seven years longer till he was unable to move either jaws, arms or legs. Became completely unable to move limbs or jaws. Twenty-one years later died, 39 years old. Autopsy showed ossification of right temporal and portion of left pterygoid, left deltoid, both pectorales major, left minor right coracobrachialis. Both biceps brachii, left triceps, right latissimus, right long dorsalis, right glutæus med. and left med. and minor, and most of thigh muscles on both sides.

Case VIII.—Reported by Caesar Hawkins, in 1844. Male, aged 22 years. First symptoms appeared at 22 years of age (June, 1843)—lumbar and dorsal regions attacked. Violently painful swellings in lumbar and dorsal regions, which disappeared, leaving in places some small bony masses. Disease later attacks neck and scapular regions. New attacks began in October, 1843, and persisted during the next six months of observation. (One year after symptoms of disease.) Lumbar vertebræ immovably bound together by osseous masses; unable to use shoulders freely, particularly on left side, owing to swelling under angle of scapula and ossification of tendon of pectoralis major. Small exostosis on rib on either side (which rib?). No congenital abnormalities mentioned.

Case IX.—Reported by G. Wilkinson, in 1846. Female, aged 21 years. First symptoms appeared at 8 months of age—stiffness in arms and lumps on back.

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Could move about and work till 11 years old, then lost motion in elbow-joints; other ossifying processes progressed rapidly. Chest cavity much diminished by curvature of dorsal vertebræ and acute angularity of ribs. Numerous thin ossific plates in anterior and posterior chest muscles, as pectorales, first dorsi and erectorides spinæ, and one plate ten and a half inches long connected to crest of left ilium and short branches running from it into rhomboidei to base of left scapula into infraspinatus; similar band from right crest of ilium to various vertebræ up to transverse process of third dorsal. Other irregular plates of bone connected spinal processes of dorsal and lumbar vertebræ and ribs connecting with sacrum by short tendinous fibres. Numerous bands of osseous matter in depressions of lower jaw; portion of bone two and one-half inches long in left biceps brachii. Two irregular bony processes, one from lower end of left humerus three inches downward into pronator teres and flexor muscles of forearm; other from outer edge posteriorly extending into triceps and extensor carpi rad. longior et brevior. No congenital abnormalities mentioned.

Case X.—Reported by King Kelburne, in 1854. Female. First symptoms appeared at 3 weeks of age—defective power of suckling and swallowing. Nourishment had to be brought into posterior part of mouth with spoon. Author discovered tumor on right side of tongue, size of large bean, judged to be of vascular nature, and therefore injected a few drops of perchloride of iron. In spite of a supposed improvement following this measure the child did not mend and died about five months later from inanition. Subsequent course of disease unknown. The post-mortem showed ankylosis (bony) between inferior maxilla and temporal bones, musc. masseter replaced by a large osseous plate descending from the inferior margin of the zygomatic process and malar bone. The osseous deposit extended as far forward as the anterior process of the malar bone, incorporated itself in its downward passage with the tuberosity of the superior maxilla, and finally amalgamated with the angle base and ramus of the inferior maxilla. These changes were on both sides. Vascular tumor on tongue had nearly entirely disappeared, leaving a fibrinous remnant of about the size of a split pea. No account given of other parts.

Case XI.—Reported by Jonathan Hutchinson, in 1860 and 1892. Male, aged 12 years. First symptoms congenital. Midwife noticed deformities; patient put in London Hospital on their account. Subsequent course of disease unknown. Left shoulder, wrist, and carpal joints ankylosed. Left humerus shorter than right and bony ridge running down outer side. Large plates of bony substance in fascia of forearm, palm and back of hand. On right side small areas of bony structure in fascia above and below elbow-joint. No congenital abnormalities mentioned. At the age of 37 years the ossifications have largely increased in size and extent, and over right scapula and in posterior fold of right axilla new bony plates have developed. Movement of thorax during respiration extremely limited.

Case XII.—Reported by William Skinner, in 1861. Male, aged 13 years. Family history negative. First symptoms appeared at 7 years of age—arms, back and right shoulder attacked. Stiffenings in arms, swellings in nape of neck and behind right shoulder. Stiffness at first was so bad that arms could not be brought to mouth. This improved later so that he could with difficulty feed himself, but hard nodules formed on different parts of body, especially breast and vertebral column. If patient receives blow or knock a hard swelling follows, preceded by pain and slight fever. Patient somewhat bent forward; slight motion in left shoulder, none in right. Arms cannot be extended and lie half bent across abdomen. Spine and scapula immovable. Small bony protuberance on ribs. Right pectorales changed into bony mass—continued into biceps until insertion at radius. Hard nodules at various places. No congenital abnormalities mentioned.

Case XIII.—Reported by Ivan Minklewitsch, in 1864. Female, aged 15 years. Family history negative. First symptoms appeared at 5 years of age—swelling

and stiffness in lower part of neck. A few years later osseous tumors in back and upper extremities began to develop, then attacked lower extremities. Three years ago pain and swelling in right knee disappeared and a week later reappeared. Exostoses in left temporal region at insert of temporal muscle; similar ones over left temporal and over acromion. Muscles forming posterior wall of right axilla ossified. Pectorales show same change, also small rough exostoses over cond. int. humeri. Left arm a similar ossification of axillary muscles. Right lower extremities: over articul. tibia tarsalis stalactite-formed exostosis, similar one over fibula; another below lower margin of patella; in soleus muscle osseous growth size of hen's egg; in fossa poplitea narrow one extending upward to thigh and in stalactite formation reaching nearly plica glutealis and inguinalis. Left lower extremities: similar growth below patella as on right side, extending upward and inward to fossa poplitea and ending at upper end of middle third of thigh; sharp-pointed exostoses on outer side of astragalus and outer malleolus. No congenital abnormalities mentioned. Report of autopsy, in 1871: New ossifications had formed at the right upper extremity on the upper posterior third of humerus and over inner part of that bone near elbow. The exostoses over cervical vertebræ and over second to fourth ribs were considerably enlarged.

Case XIV.—Reported by Johanne Zollinger, in 1867, and Theodor Billroth, in 1869. Male, aged 24 years. Believed to have been caused by fall (?). First symptoms appeared at 3 years of age—arms first showed stiffness in movements. Movements of arms grew more and more difficult, and after a few years present state developed. Bent forward in lumbar position of vertebral column. Head bent forward; mouth can only be slightly opened for introduction of little finger. Both sternocleidi muscles changed to rigid cords. Arms in slight abduction and flexion in elbow-joints; tendons of pectorales and deltoids hard and very tense. Back shows many protuberances of hard osseous consistency, situated mostly around the lower part of lumbar region; they branch off in many different directions. Especially large is a ridge going on either side into glutæus maximus; others in regions of sacrospinalis, irregular and asymmetrical. Both latiss. dorsi nearly entirely converted into bone. First interphalangeal joint of left middle finger thickened by exostoses, and small roundish exostosis on left index finger. Small exostosis at inner side of left tibia and on outer side of r. cond. ext. femur. No congenital abnormalities mentioned.

Case XV.—Reported by Muenchmeyer, in 1868, and A. Jurasz, in 1873. Female, aged 22 years. Family history negative. Fall when 4 or 5 years of age. First symptoms appeared between 4 and 5 years of age—neck and right arm; nodules on nape of neck. Development of right-sided scoliosis; stiffness of neck and right arm. After first nodules on nape of neck had disappeared others came in different parts of body. Brain fever at 12 years of age. In May, 1864, had scarlatina. Patient does not recollect ever having been able to turn her head freely or move arms away from her sides. Asserts never to have been able to dress herself or to do up her hair. Cervical column and head completely immovable; position of head in right-sided caput obstipum. Dorsal spine right-sided scoliotic and immovable; both scapulæ firmly fixed. Left arm nearly completely immovable in elbow-joint; a nodulous osseous mass between cond. ext. hum. and olecr.; left biceps brachii one solid mass and long bony stalactite extending from middle toward axilla. Hard ossified parts also in deltoid. Both latiss. dorsi and trapezii, teres major and minor are changed to osseous masses. Also the enormously thickened lig. nuchæ. No congenital abnormalities mentioned. Latest observation in 1869; all changes have remained, and in addition the whole back one solid mass, protuberances, nodules and ridges, with some softer parts intermixed. A six-centimeter broad osseous mass leading from vertebral column to scapula fixing the latter completely. On the supraclavicular fossa everything except the omohyoideus seems to be ossified. In right leg whole gluteal mass ossified; from thigh down to knee several broad bony masses. During time of observation various places all over the body showed swellings, which disappeared again.

Immovability of jaws increased so that patient could take only liquid food. In 1869 disease got worse and continued from bad to worse. In 1872 m. gracilis involved. In November mental trouble began. February 26, 1873, she fell and broke her right arm. Plaster-of-Paris dressing, cured in two weeks. No excessive callus. Died in thirty-second year of her life. Mays' autopsy report in 1874.* The autopsy report by Mays is too lengthy to be given here. It adds some exostoses to the ossifications found during lifetime, but shows the cerv. muscles not to have been ossified, only hard and infiltrated with a connective tissue hyperplasia. No examination of fingers or toes. The spinal cord and periph. nerves were examined by Doctor Schultze† and found normal.

Case XVI.—Reported by Breschet, in 1869. Skeleton. Subsequent course of disease unknown. In addition to real exostoses, also multiple and symmetrical ossifications of muscular attachments. Skeleton in Musée Dupuytren (Paris).

Case XVII.—Reported by William Byers, in 1870. Male, aged 17 years. First symptoms appeared at 8 months of age—small tumors size of small marbles appearing in different parts of body. Various parts of body attacked. Stiffness of joints, hips and shoulder-joints. The first nodules disappeared, but after twelve to eighteen months stiffness of joints set in again. Gradual ankylosis followed, and at the age of 10 years left hip and shoulder joints were completely ankylosed. Later the muscular system began to show ossification. Chest became as though enclosed in a complete sheet of bone, leaving no trace of outline of ribs. Head was immovably fixed by ossification of both sternocleides. Up to his seventeenth year muscles of mastication remained unaffected. Could at first move slowly and cautiously over smooth surface; with ankylosis progressing this became impossible. No congenital abnormalities mentioned. Remained small; sexual infantilism. Died at the age of 21 years; muscular system almost completely ossified.

Case XVIII.—Reported by Florschuetz, in 1873, and Gerber, in 1875. Male, aged 12 years. Family history negative. First symptoms appeared at 5 years of age—hard inflamed nodules in right latiss. dorsi below right scapula. Latiss. of left side, upper extremities and back attacked later. After having attacked shoulders and upper extremities it progressed to muscles of back, trunk, neck and mastication (tempor. and masseter) and upper parts of lower extremities. Frequent nocturnal epileptic attacks. Absolute stiffness of large muscular groups. The greater part of muscles of neck, all muscles of mastication, causing complete lockjaw (nutrition through gap in teeth), muscles of back, breast and arms (arms are immovably fixed to sides of body, only left forearm functions slightly), and finally those of left thigh and hip and part of right thigh are ossified. Many muscles which are not ossified are atrophic, evidently in transitory stage, previous to ossification. No congenital abnormalities mentioned. Penis normal in size relative to age. Scrotum atrophied and testes quite undeveloped. Autopsy report, Mays,* 1874. Very extensive autopsy in which several exostoses were found and most of the ossification diagnosed during life confirmed except in the cerv. muscles, especially in sternocleidi mast. and the delt. and adjoining extensor musc. of the arm, which only showed a firm infiltration, together with development of the connect. tissue hyperplasia. Fingers and toes not examined. Spinal cord and periph. nerves found normal by Schultze.†

Case XIX.—Reported by Edward F. Hamilton, in 1874. Female, aged 30 years (?). No clinical history obtainable. Subsequent course of disease unknown. Body brought to Medical School of St. Stephen's Hospital. Body inclined to left side; head bent

* Virchow's Arch. f. patholog. Anat. und Phys., bd. 74, p. 145.

† Erb and Schultze; ein Fall v. progress. Muskelatrophie, etc. Arch. f. Psychiatric und Nervkrkhtn, 1879, bd. 60, h. 2, pp. 385-6.

* Op. cit.

† Op. cit.

forward, arms closely applied to sides; forearms flexed and pronated. No motion of shoulder-joints nor of scapula upon trunk. Left side of thorax contracted; right ilium nearly in contact with ribs. Lower limbs flexed and adducted, right leg and foot much inverted; ankle joints stiff. Skeleton showed: Along anterior border of left masseter a ridge of bone preventing any movement of jaw. Head immovably joined to upper cervical vertebræ. Spinal column forms one solid mass; cervical, dorsal and lumbar vertebræ firmly joined by copious bone deposits; left shoulder-joint perfectly stiff and large plate of bone fixed to humerus near insert. of pector. maj. reached up to coracoid proc., then down and inward till identified with costal cartilage. Connecting the shaft of right femur and trochanter immense osseous growths resembling stalactites had formed, branched into the fibres of extensor muscles. Similar growths were upon dorsum of ilium. Osseous deposits at patellas, insert. of quadriceps, extensor, tendon and many ligaments of soles of feet had ossified. Between muscles of back from occipital bone to inferior angles of scapula and down to crest of ilium. Consolidating hip-joints were enormous plates of ossific matter; in front of left femur osseous deposits bearing resemblance to a second femur. This deposit had been formed in cellular tissue between rectus femoris and cruræus.

Case XX.—Reported by Bennett, in 1874. Female, aged 11 years. Family history negative. First symptoms appeared in infancy—swellings over shoulder and back of neck. Irregular attacks over various parts of body. On right side from protuberantia occipit. to supraspinous fossa a bony growth is stretched, apparently situated in trapezius, irregular masses of bone are deposited in location of rhomboid muscles, and bony processes branch upward and inward from latis. dorsi, portions covering scapular angles, other branches going down from those to crest of os ilium. Across middle and upper parts of loin transverse osseous bars have developed following the course and fibres of the muscle. A spur has commenced to grow from either ulna near attachment of pronator quadratus. No congenital abnormalities mentioned. Cast of girl demonstrated.

Case XXI.—Reported by Dittmeyer and Gerber, in 1875. Female, aged 8 years. Family history negative. First symptoms appeared at 6 years of age—complained of pains in right side of nape of neck and shoulder. Stiffness of neck and right shoulder; some difficulty in breathing. Right shoulder grew immovable, breathing became more difficult. The left shoulder stood higher, head had been drawn toward left side. Right trapezius muscle stiff and hard, so that the head could not be moved to either side. The supraspinati and teres minor grew as hard as a board and arms were tightly drawn to sides of thorax. In serratus post. super. one could feel the daily spread of the ossifications along the separate muscles. All other muscles were free. No congenital abnormalities mentioned. General health, with exception of the difficult respiration, was good; all organs functioned well. Autopsy report, Mays.

Case XXII.—Reported by V. P. Gibney, in 1875. Female, aged 10 years. Family history negative. Attack of diphtheria (?). First symptoms appeared at 10 years of age—had been perfectly well up to attack of diphtheria. Subsequent course of disease unknown. Muscles involved were latiss. dorsi, scaleni and erector spinæ. Right arm was held down by tendon of latiss. dorsi. Also lateral curvature. No congenital abnormalities mentioned. No mention of microdactylia. At suggestion of a colleague acid. lact. was given, with no amelioration of symptoms; but there was no further progress during the next year and a half of observation. Seen again in 1893. Had been working in a millinery shop since 1884. No change of status. (Doubtful case.)

Case XXIII.—Reported by Huth, in 1876. Male, aged 4 years. Family history negative. First symptoms appeared at about 2 years of age—hard spots under chin. Gradual extensions toward angles of lower jaw; then chest and other parts of body. Hard swelling size of egg developed over middle of sternum at about 2

years of age. It disappeared again. On shoulder and both upper arms swelling and hardness developed. In the spring of 1874 a swelling appeared on forehead, extending over eyelids, eyes being entirely closed. This disappeared again. Upper arms lying closely to trunk and fixed; lower arms movable. Head, neck and thorax muscles hard and stiff like armor. Both glutæi, especially of left thigh and leg, ossified. In abdominal muscles a cord thick as finger running from angle of ribs to os pubis. No congenital abnormalities recorded.

Case XXIV.—Reported by Nicoladoni, in 1878. Female, aged 7 years. First symptoms appeared the first year of life—stiffness in muscles of neck. Later stiffness in muscles of back. At present two hard ridges corresponding to sacrolumbales. The scapular muscles are ossified so that the shoulder-blades are immovably fixed to thorax. At upper part of biceps brachii are small bony plates in various parts of the muscle, its tendons changed to hard fibrous cords. The same changes present in sternocleidomastoidei. The contracted right knee-joint can only be extended to 120°. Semitend. and membranos. changed to two hard lumps. Pectorales also ossified and the axillary fossæ are bounded in front and posteriorly by rigid walls. The muscles of the lower jaw are also involved. No congenital abnormalities recorded.

Case XXV.—Reported by H. Helferich, in 1879 and 1883; Carl Mannz, in 1893, and Arthur Manneberg, in 1896. Male, aged 16 years. Family history neg. First symptoms appeared at 6 years of age—numerous hard nodules appeared upon head and gradually disappeared again. Later similar painful nodules appeared on back; arms and back became stiff. Between the twelfth and thirteenth years slow development of closure of jaws; first right molar had to be extracted to allow feeding. When 15 years old could still push out tongue between teeth. Jaws now firmly closed. Arms gradually became stiff and nodules developed all over them. Axillary fold hard; masseteric region hard ridges from the process. zygomatic to lower jaws. Jaws locked. Neck in region of hyoid bone hard, but bone and larynx movable. Thorax, pector. maj. shows ossified swellings. Back, swellings of osseous hardness spreading like antlers from pelvis upward. Right hip immovable; left hip normal; left gluteal region, flat resistant bone-hard mass; abscess developed in right inguinal region. First case where attention was drawn to microdactylia of thumbs and big toes in this disease, although anatomical defect was erroneously thought to be defect of an entire phalanx instead of smallness of metacarpal or metatarsal bone. In later observation (1887) it is reported that case had remained fairly stationary since first observation. In latest observation (1896) by Manneberg, quite an additional progress is noted, comprising old and new muscular groups.

Case XXVI.—Reported by Partsch, in 1882. Male, aged 17½ years. Family history neg. First symptoms appeared at 10 years of age—swelling after fall on right shoulder; disappeared again, but hardness remained in right side of neck. Developed stiffness of muscles, which affected right shoulder and upper arm; later left arm; after fall on and wounding forehead a swelling of os frontale followed. Two years ago muscles of left side of neck hardened and drew head over to left side. About same time ossification of masticating muscles set in so that mouth can only be opened 1 cm. Head bent to left and fixed forward; both arms flexed in elbow-joints, right cannot be approached to body, left only with great exertion. Both shoulders fixed. Cervical column rigid. Sternocleido fibrous, but no bony enclosures. Scaleni show osseous changes. Right scapula carries a small roundish osseous tumor 4 cm. long, 2 cm. broad; lower border of left latiss. dorsi entirely osseous. Left brach. ant. contains osseous growth. Right latiss. dorsi shows osseous growths extending to upper arm and triceps. In back: two osseous plates (11 cm. long), one in each fascia lumbodorsalis. Exostoses 1½ cm. long near right spin. ant. os. il. Broad bony plate in right tens. fas. lat. and narrower one in right

sartorius. Similar one 3 cm. broad and 13 cm. long in left sartorius. Microdactylia of thumbs and big toes.

Case XXVII.—Reported by Herm. Kuemmel, in 1883. Male, aged 13 years. Family history neg. First symptoms appeared fourteen days after birth (congenital)—foster-mother then observed striking deformity of spine and back and restricted mobility of both arms, which could only be slightly abducted from thorax. In second year various fluctuating swellings were observed upon back. They shrank and hardened into exostoses. Except some hardening of more swellings no particular change occurred until twelfth year, when gradual contraction of left knee-joint began. Head inclined forward and slightly to right side; movements very restricted. Small exostosis on occiput. Arms hardly movable from sides of thorax; upper part of body bent forward and curved. Floor of oral cavity forms callous mass, including os hyoid, and thyroid cartilage. Both sternocleidi and sterno-hyoid fibrous. Entire cervical spine bent forward and forms one continuous solid mass. Ligam. nuchæ much thickened. Costal cartilage forms pectus carinat. In both pectorales major, bony bars extending in right to coracobrachialis. Both axillary posterior walls contain bony bars. In right rect. abdomin. tough fibrous and thin bony band. Both psoas tough callous masses. Dorsal scoliosis to right, lumbar to left. From seventh dorsal vertebra downward processi spinos. form one solid osseous mass. Both large trochanters broadened and prominent. Muscles of left thigh posteriorly changed into tough callous mass, with freely movable bony protuberance. Left leg flexed, angle of 45 degrees. Both thumbs nearly (?) normal; show bony ankylosis in interphalangeal joints. Microdactylia of both big toes ascribed to absence of first phalanx. (Real cause is probably smallness of metatarsal bone.)

Case XXVIII.—Reported by Uhde and Pinter, in 1883. Male, aged 16 years. Family history neg. First symptoms appeared first year of life—swellings on head, which disappeared after some time. Patient broke arm in 1876. Fracture healed perfectly and shows *hardly any trace* of callus. Head and shoulder joints bent forward; arms ankylotic in shoulder joints; movements in elbow joints limited only. Ankylosis of interphalangeal joints in both thumbs. Microdactylia of both great toes (with same probable error as above).

Case XXIX.—Reported by Krause and Pinter, in 1883. Female, aged 11 years. Family history neg. First symptoms appeared soon after birth—swellings in several places on scalp, which disappeared again after several months. At age of 4 years swelling in some of the deeper muscles of that region. It spread rapidly, then decreased, but left board-like hardness, fixing scapula firmly to thorax. Short time after same process occurred on right side. Disease progressed to left latiss. dorsi and biceps brachii, part of intercostals and serratus. Two years ago right mylohyoid, geniohyoid and part of biventer followed with swellings, which, disappearing, always left board-like hardness with spinous growths 1.38 cm. tall; circumference at mammilla 62 cm. Slight kyphosis, convexity at seventh dorsal. Left shoulder completely stiffened. Deltoid atrophic. Biceps ossified, fixing elbow in slight flexion. Broad band from spine to angle of scapula, two larger on right. Over scapula and thorax flat exostoses. Left latiss. dorsi totally ossified. Ribs below fourth rigid and unyielding. No mention of microdactylia.

Case XXX.—Reported by Gyula Pinter, in 1884. Female, aged 20 years. Family history neg. First symptoms appeared at 4 years of age—hard nodules on back and disturbances of mobility of both shoulder joints. Patient does not remember ever having been able to lift arms above shoulders. At 6 years abduction of arms became so restricted that she could not write when seated at school desk. Other joints remained free until twelfth year. About the twelfth year, during formation and healing of suppurating swelling over right tibia, nearly all her joints lost greater part of mobility. In 1881 another suppurating swelling appeared on left leg, with a similar

injurious effect on the various joints, rendering them still more rigid. When lying in bed rigidity of body like that in tetanus. Head; caput obstipum nearly immovable; chin held out and downward; ankylosis of lower jaw-bone; only lateral excursion of about 2 mm. in horizontal plane possible. Both temporales and masseters of bony hardness. Thorax absolutely immovable during respiration, owing to complete ossification of both pectorales major and minor. Axillary fossæ rigid walls; both cucullaris ossified. A bony ridge 2 cm. broad, extending in middle line of back from prominentia occipit to seventh cerv. vert. Back and arms, two bony ones on either side of ninth and tenth dorsal vert. Left 10 cm. long, 2½ cm. wide, sends branches toward left scapular angle and down to crista ilei. Right, 6 cm. long and 2 cm. wide, sends similar branches anastomosing with other side. All firmly united with underlying parts of skeleton. Shoulder-blades immovably fixed to trunk. Stalactites of bone in ant. and post. muscles of both upper arms, and in left pronator teres and radial ext. Pelvis and legs, exostoses 3 cm. long on both crist. near ant. sup. spine. Hip joints immovable; muscles origins from both tuber. and ram. asc. ischii and trochant. maj. all ossified. Bony ridges in lower halves of bic. fem. and semitend. and membr.; muscles of lower legs and feet free. Microdactylia of both big toes.

Case XXXI.—Reported by O. Kohts, in 1884. Male, aged 23 years. Family history neg. First symptoms congenital. Could not move since his ninth year, having then made his first attempt to walk. Severe pains ensued after he first tried to walk, which made him take to his bed until his fourteenth year, when he entered the hospital. Patient 141 cm. tall. In bed could not change his position. Head and trunk slightly bent forward and to right. Lower jaws ankylosed. Masseter and pterygoidei apparently ossified. Exostosis 2 cm. long on inframaxillary bone. Ligam. nuchæ and muscles cucullar. and splen. partly ossified. No thoracic respir. movement. Both clavicles show large exostoses. Shoulder joints ankylosed. Both maj. and minor pectorales contain bony plates; entire lower margins ossified. Both arms firmly fixed to thorax. Both coracoid muscles and left brach. int. ossified. Right elbow-joint ankylotic. At fourth rib exostosis 2 cm. long. Left elbow limited mobility. Muscles coming from right condyl. int. humeri ossified in upper parts. Both hip-joints ankylotic. Deep glutæi and insert. of right quadrat. ossified. Left tens. fasciæ and tendon of right biceps femur and insertion of right gastrocnemius contain bony plates. Spinal column scoliotic; vertebr. column ankylotic; muscles along both sides atrophic and ossified. No mention of microdactylia.

Case XXXII.—Reported by T. Sympson, in 1886, and Stonham, in 1892. Male, aged 6 years. Father rheumatic and presents same congenital deformity of toes as son. Fell on shoulder when 5 years old. First symptoms appeared at 5 years of age—painful swelling formed on right shoulder after fall one week earlier, then shrank gradually; three months later a similar swelling appeared on left scapula. Various swellings formed and disappeared again on back and chest. Jan. 16 (at demonstration) saddle of bone over loins, making stooping impossible. Margin of each latiss. dorsi, of each teres major and long head of r. triceps were occupied by a series of lumps apparently bony; similar lumps present a little below occiput in left trapezius, the middle of neck and on right side, and on supraspinous fossæ. Six years later (1892) two bones size of filberts found on inner side of post. sup. spine of r. ilium. Patient fell Aug., 1887, and broke both bones of left forearm. Fracture united well. A firm hard swelling involving left vastus ext. and int., extending from just above patella upward for about four inches, consequent to a fall upon the knee. A similar swelling posteriorly in the popliteal space, biceps and semitendin. Motion of knee limited. A nodule size of filbert in middle of inner side of left thigh, situated in sartorius muscle. Feet show head of each metatarsal bone large and prominent; big toe on either side small and apparently consists of only one phalanx (?) and is directed toward outer border of foot. (No X-ray.)

Case XXXIII.—Reported by Willett, in 1886, and Stonham, in 1892. Male, aged 4 years. Family history neg. First symptoms appeared at 6 months of age—swelling at superior angle of right scapula. When 1 year old swelling along vertebral border and inferior angle of right scapula. Six months later prominence noted in left erector spinæ and a little later a swelling near lower angle of left scapula. Right frontal eminence enlarged. Hard growth over left transverse processes of fourth and fifth cervical vertebræ. Right lateral dorsal curvature of spine, with slight compensat. curvature in lumbar region to left. Slight kyphosis of upper dorsal spine. Both scapulæ fixed. Hard growth along vertebral border of right scapula. Latiss. dorsi at scapular angle and in its tendinous portion ossified. A similar, less extensive hardness on left scapula and latiss. dorsi. A bony nodule on front angle of eighth rib, similar on left. Both erectores spinæ ossified. On right knee just above head of fibula hard mass 1 inch in diameter, with nodule size of pea. Inner tuberosity of right tibia slightly thickened. Both big toes hallux valgus position, displaced outward and under second toe. Shortened microdactylia, only ungual phalanx present on each side (?). In Stonham's subsequent report six years later no correction of this statement of a very unusual condition is made.

Case XXXIV.—Reported by Rickman J. Godlee, in 1886. Male, aged three years. Family history neg. First symptoms were noted when 1 year old, but may have been present long before, as nothing was learned of previous history from parents—bony plates in right latiss. dorsi and a nodule near right knee. Was not seen again till two years later, when present status was noted. In r. latiss. dorsi along outer border and post. fold of axilla, irregular bony mass; arm cannot be abducted more than sixty degrees, but can be completely adducted. Flexion of shoulders considerably limited; extension less; rotation inward more than outward. On left side post. a large elastic mass extending medially to nearly within the middle line and considerably below scapula, and upwards over shoulder to near clavicle. Scapula appears fixed in mass below angle. This mass had disappeared on March 23, 1886, but there is a hard mobile mass below left angle. Similar hard bodies, apparently bony, not movable, on free edge of left latiss. Seen again May 20, 1886; over the fourth rib, behind in post. axillary, ossifications. Over third rib in posterior axillary line, similar ossifications. Shoulder cannot be raised to right angle. Head drawn down to left shoulder. Some hard masses in muscles on outer side of neck (trapez. and right mast. excepted). Left arm cannot be raised higher than 45 degrees from trunk; scapula firm. In post. axillary fold hard nodulous mass. From angle of scapula a hard crest runs over to first lumbar vert. Back strongly curved; lower left ribs down on crest of ilium. Pollex valgus and microdactylia of thumbs. Hallux valgus and microdactylia of toes.

Case XXXV.—Reported by R. v. Volkmann, in 1887. Male, age (?) Skeleton. Subsequent course of disease unknown. Various parts of his muscular system had been partially substituted by bony masses, which had grown into them. V. Volkmann had made attempts by removing some of them to restore a certain movability of ankylotic and immovable parts of the skeleton. The only published mention of this patient is made in a brief and vague reference in a discussion of Helferich's' case. The extirpated bony masses showed at one end an epiphysial layer of hyaline cartilage.

Case XXXVI.—Reported by Alfred Austin London, in 1887. Male, aged 43½ years. Family history neg. Accuses slight blows when 10 years old. First symptoms appeared at 10 years of age—joints of legs and shoulders stiffened. Gradually until his thirtieth year grew quite stiff and helpless. Jaws could only be opened very little. Over sacrum a bedsore formed from which pieces of bone were constantly exfoliating. Ossification of deltoid from acromial origin to insertion; also part of coracobrachialis. Scapulæ immovable on trunk, their inferior angles soldered by dorsal buttress of bone attached by seventh to ninth ribs. Latiss. dorsi ossified nearly entire lengths. Ossification of left brachialis anticus, and bony rims around both elbow-joints. Sacro-iliac ligament ossified. Both hip-joints ossified. Bone rods developed in lower

part of glut. max. and extending from sacrum to femoral insertion. Huge bony projection on back of femur. Dorsal lumbar curve, with slight rotation to right. Very rigid, due to ossification of the capsular ligaments of the articular processes and many of the supraspinous ligaments. Died at age of 46. Microdactylia of both big toes. (No mention of thumbs.) General synostosis of the neural arches. All the ribs except the eleventh and twelfth are ankylosed to vertebral column. Bones have undergone eccentric atrophy and are consequently rather light.

Case XXXVII (?).—Reported by E. Schwarz and Cl. Eichhorst, in 1888. Male, aged 40 years. Family history neg. First symptoms appeared at 39 years of age—a hard mass in posterior muscles of thigh. Subsequent course of disease unknown. Exostoses also on right humerus and on right femur in the region of the trochanter minor. Hyperostosis of right fibula. Suffers from tabes dorsalis. This case, as well as another from Eichhorst's clinic, both affected with spinal diseases. Do not belong to myositis ossificans progressiva. No microdactylia.

Case XXXVIII.—Reported by Kronecker, in 1889. Male, age (?) First symptoms appeared in (?) later years—first pains in nape of neck and in chest, two years previously. In November of the previous year the scaleni became affected. Since one year dyspnœa increasing; the cucullares are involved and the process is evidently progressing. Patient has pains in neck and one feels distinctly the increasing ossification. No microdactylia. (Doubtful, atypical case.)

Case XXXIX.—Reported by J. v. Bokai, in 1899. Male, aged 5 years. Family history negative. Rickets. First symptoms appeared at 4½ years of age—nape of neck and back. Subsequent course of disease unknown. Muscles of nape of neck, back and serrat. anticus maj., pectorales maj., cucullaris and latiss. dorsi are affected. No congenital abnormalities recorded.

Case XL.—Reported by Ivar Svensson (Sabbatsberg Hospital), in 1891. Male, aged 14 years. Family history neg. Fell when 4 years old (?). Age of first symptoms unknown—stiffness of body, arms and back. Stiffness has been gradually increasing. Stiffness of head and back and arms. Scoliosis on nape of neck; these are hardened and flattened and atrophied about the level of the levator anguli scapulæ, with fixed shoulder-blades; arms cannot be raised more than one-fifth of right angle from thorax. Otherwise arms can be lifted to horizontal plane, but not higher. Passive efforts to overstep this give crepitation and pain. In the height of axillary fossa a flat piece of bone of about 7 cm. length and 2 cm. width intimately connected with the tendons of latiss. dorsi and teres major and to the edge of the scapula. At the inner sides of both scapulæ parallel with spine one can palpate a round bony formation of 5 cm. length and hardly 1 cm. width. No mention of microdactylia in either fingers or toes.

Case XLI.—Reported by C. Studsgaard, in 1891. Female, aged 4 years. Family history neg. Measles and whooping cough when 2 years of age. First symptoms appeared at 2 years of age—noticed swelling after measles and whooping cough. (Where?) First swelling (where?) disappeared again during use of cod-liver oil. Growth of bony consistency appeared after another year without any cause on the left side of the neck, rendering movements of the neck, especially downward, very difficult. Diffuse osseous swelling appeared on left lower jaw like convex continuation, cylindriform, size of finger, from base of that bone in region of premolar to incisura semilunaris sterni, and firmly attached to left horn of the corpus ossi hyoidei with one end, with the lower movably connected with sternum. Slight movements of maxilla are still possible, also small side motions of the head. The mass is situated anatomically as if it were as ossification of the sternohyoid muscle, wherefrom the bony part continues to the lower jaw. Bone of neck extirpated February 4, 1891. Dismissed March 20, 1891. New bone formation on location of extirpated bone from neck. Microdactylia of both thumbs and large toes and ankyloses of their phalanges.

Case XLII.—Reported by R. Gordon McDonald, in 1891. Female, aged 4 years. Family history negative. First symptoms appeared at 2 years of age—lumps appeared on left side of neck about middle of sternocleido. Later on lumps appeared on forehead and back (dorsal vertebræ). All lumps disappeared after some time. Head fixed; unable to move it; sternomastoid, trapezius, stylohyoid, omohyoid and sternohyoid ossified. Over left frontal eminence a large node; smaller one over left temporal bone and right border of occipital. Both elbows can be moved only eight inches from trunk; teres major, latiss. dorsi, edge of pectorales major becoming gradually ossified. Scapulæ fixed to ribs, right less than left. At inferior angle of both scapulæ large nodes; on left superior angle a similar one; right side free. Various-sized nodes along the vertebral spinæ from head to sacrum, also along lateral parts of several ribs and over crest of ilium. The superficial muscles of back are gradually ossifying. Ribs are also becoming fixed and she is unable to take deep inspiration. Teres major was, experimentis causa, exsected during hospital stay. It was totally ossified. Result as to mobility—none. No congenital deformity mentioned.

Case XLIII.—Reported by Bilton Pollard, in 1892. Male, aged 9 years. Family history neg. First symptoms appeared 6 months of age—lumps formed on lower angle of right scapula and nodules appeared on boy's ribs. In second year neck was getting stiff. In third year lumps formed between right iliac crest and the last rib. In his fourth year hard nodule formed a little below patella after fall on right knee. New lumps formed, his left arm stiffened and bony bands appeared in his neck up to his seventh year, but none during his seventh, eighth and ninth years. Neck quite stiff; rotation and raising of head barely possible. Bony band between chin and sternum extending from the lower border and from the right of lower jaw to right side of cricoid cartilage, there dividing into two, the right ending just above sterno-clavicul. articulation, left above clavicle between the two heads of left sternocleido. R. arm can be raised to angle of 45 degrees only together with scapula. Axillary folds contracted. Scar there from bony operation five years ago. Beneath it bony plate slightly movable, apparently in latiss. dorsi. Pectorales and deltoid hardened; hard mass on border of radii two inches above styloid process. L. arm equally stiff. In course of teres major runs thick band of bone, extending free from inferior scapula angle almost to humerus. Bony projection at left-elbow bend connected with biceps tend.; another mass a little deeper down involving brach. antic. Spine very rigid, and on both its sides bony nodules and plates in dorsal and lumbar regions. On outer and post. part of r. thigh flat osseous plate; smaller plate on left. On each thigh strong bony spiculæ, extending from adductors tubers. upward to tend. of adduct. magnus for about two inches. Irregular-shaped movable plate between lower fold of left condyle of femur and patella and a smaller one near outer tuberosity of tibia. Inner borders of both tibiæ show bony growths just below tuberosities. No congenital abnormalities mentioned. Pieces of bone were exsected to improve mobility of arm and neck, but they soon formed again, and the slight improvement gained by the operation was lost.

Case XLIV.—Reported by Luigi Bernacchi, in 1892. Male, aged 7 years. Family history neg. First symptoms appeared at 3 years of age—muscles stiffened in nape of neck without inflammatory symptoms. Small hard tumors formed in right cucullaris over tuberosities of frontal and parietal bones. About a year ago stiffness of humero-scapular joint appeared. Left arm could not be raised above horizontal line. New hard osseous growths have formed along the spinous processes of dorsal vertebræ, and growths in lumbar muscle so as to form line from neck to os sacrum. Two cervical projections over frontal tubera. Head slightly inclined forward and to the left; movements restricted. *Neck and back;* from the occipital insertion of right trapezius near middle, irregularly formed osseous tumors forming one solid mass of muscles. *Thorax and abdomen normal;* small exostoses above styloid process of radius. Posterior wall of axilla shows fibrous consistency and is shorter than normal.

The scapulæ are united by bony ridge about their middle; an osseous tumor within the dorsalis major can be felt, size of thumb and somewhat movable. No mention of microdactylia. (Photograph of boy shows indication of microdactylia of thumbs.) No mention made in text.

Case XLV.—Reported by Ludwig Rabek, in 1892. Female, aged 3½ years. Family history neg. First symptoms appeared at 6 months of age—hard nodules in region of shoulder-blades. Parents noticed movements of upper extremities were restricted; movements of lower jaw have been impeded since one year before. Difficulty and restriction of movements of lower jaw and both upper extremities. The distance ad maximum between the front rows of teeth is only 0.5 cm. Right masseter hardened and hypertrophied. The arms cannot be raised to horizontal line. Right elbow-joint contracted, extension incomplete, flexion normal. Shoulder-blades only slightly movable. Osseous tumors arise on many parts of the body, mostly on back. Large one in right axilla extends to latiss. dorsi as far as scapula line of tenth rib. Another on left side reaches eighth rib. There are four more in the scapula and one in lumbar region, all in the muscles. Right biceps changed into hard movable mass united with shoulder muscles. Small pea-sized nodules in right pectoral. Big toes of both feet contracted outward and downward so that each lies under the second toe. (Microdactylia and hallux valgus.) Congenital deformity.

Case XLVI.—Reported by J. Brennsohn, in 1892. Male, aged 20 years. Family history neg. First symptoms appeared in early childhood—a certain clumsiness of motion and elevation of right shoulder. Later a scoliosis, and three years ago right arm grew stiff, then left arm, then the trunk and subsequently the legs. Had to give up work in factory. The muscles of the posterior cervical region are completely ossified; all the anterior muscles feel rigid, especially the scaleni and right sterno-cleido; below the right lower jaw the mylohyoid forms a bony growth like a stalactite. Mobility of head nominal; shoulders quite stiff; arms cannot be abducted from side of thorax. Left elbow-joint ankylotic; right only partly movable. Both anterior and poster. walls of axillæ ossified; also deltoids, serrati antici maj. and intercostales. The lower parts of left biceps and brachialis indurated and tense. Hand- and finger-joints free. Ramified osseous growths from both cristæ ossis ilii into both glutæi max. Some mushroom-like exostoses from left crista ossis ilii. Both big toes micro-dactylic (metatarsus primus?) and in valgus position. Brennsohn ascribes it to missing of basal phalanx. No X-ray taken.

Case XLVII.—Reported by V. P. Gibney, in 1893. Male, aged 10 years. Family history neg. First symptoms appeared at 5 years of age—was admitted to the hospital at this time to sever a strip of ossified muscle, but was removed by parents before operation. Subsequent course of disease unknown. Head tilted to right; upper extremities bowed so that thumbs touch elbows of opposite side; elbows abducted from trunk; shoulders stiff; right clavicle has extra curve at outer half; greatest convexity posteriorly. Left clavicle curved entire extent; convexity toward neck. From its middle springs irregular bony mass elevated half an inch. Base of this mass spreads in clavicular portion of pectoral. major and continues in its pectoral part, terminating in anterior wall of axillary space. Over sternal articulation of second rib small exostoses tapering off into pectorales mass. Similar ones over sternal end of third and fifth ribs. None on right; about middle of fifth rib, just in front of axilla, a bony enlargement extending back and downward, involving the whole area of latiss. dorsi and serratus magnus. March, 1893; the bony tendon of the right latiss. dorsi was divided and a piece of bone about one inch wide was excised, but new bone was thrown out and rendered void the effect of the operation. An osseous tumor the size of a peanut over tendo achillis which was removed did not return. No mention of microdactylia. Private advice kindly given me says that author failed to take notes about presence or absence of microdactylia.

Case XLVIII.—Reported by R. Virchow, in 1894, and Bollinger, Linsmeyer, Kraske, Wollenberg, A. Weil, Nissim and Weil, Faulkner, Lyon, De la Camp, Ponfick, Birch-Hirschfeld. Male, aged 29 years. Family history neg. First symptoms appeared at 18 years of age—swelling and pain in maxillary articulation. Pain, swelling and final ankylosis of maxillary and various joints of extremities. Osseous masses on both sides of median line of neck. Bony elongation from right process coracoideus extending along tendon of that muscle and of the short head of the biceps for about 5 cm.; somewhat less on the left side. Ankylosis of right shoulder-joint. Ossification of right triceps extending upward to teres and dorsalis muscles, also some ossification in anconeus. Small cartilaginous nodules on the tendons of abductor longus and extensor brevis pollicis. The left arm shows the same changes as the right and a cartilaginous, fork-like formation over elbow-joint. Irregular osseous masses in upper part of trapezius and over lumbar region projecting at various angles and of varying sizes. Bony formations project from the sacro-iliac articulation to the right and left sides, reappearing in the iliac fossæ and extending to the coxofemoral joints, which they immobilize, reaching to the great trochanter and into the muscular insertions thereon. Thickening of tuberosity of right tibia; exostosis of head of astragalus. Heads of metatarsal bones thickened as in arthritis deformans. Left leg shows similar changes as right one; osseous mass extending from popliteal fossa down between the two bones and immobilizing knee-joints. Thickening and ankyloses of ankle-joints. Ankyloses of big toes; from the tip a long bony appendix is branching off, size of a walnut, like a supplementary toe. In the discussion of this case Gerhard drew attention to the microdactylia of a big right toe. Case began rather late in life, but except this one symptom, has all the earmarks of the disease. The epiphyses of the long bones are so soft that pins can be pushed into them.

Case XLIX.—Reported by A. A. Kisel, in 1893, 1906-1909. Male, aged 1 year and 7 months (13 years?—16 years?). Family history negative. First symptoms appeared at 1 year of age—swelling in nape of neck size of small walnut; no accompanying pain or inflammatory symptoms. After three or four weeks most of tumors disappeared without leaving any trace. Some others seemed to soften and discharged a puriform liquid. All without symptoms of inflammation. *Later,* head has been drawn forward and posterior part of neck has become very rigid and quite immovable. Impossible to turn head backward. Head drawn forward. Chin touches thorax. Motions of head very limited. The entire posterior part of neck appears considerably swollen; the borders of the muscles very much thickened. The sternomastoid and pectoral muscles present some changes. Movements of right shoulder very limited, especially abduction and elevation. April 25, 1893, a new tumor appears in the right flank, with the same character as the others and reaching size of a small walnut. When 16 years old bone-like hardening of ant. and post. cerv. muscles, muscles of thorax, scapulæ, shoulders, abdomen, osteal attachment between twelfth rib and crista ilii; exostoses on lower jaw and scapular spine; head inclined forward and to the left, and immovable; head, vertebræ, neck, thorax and upper extremities in shoulders all one mass of bone; limited movement in elbow-joint. Improvement at first under K. J. At 13 years walked well, but now unable to leave bed. Extirpation of small tumors; tumor shows muscle cells much swollen, reddish-yellow and œdematous. Microscopical examination; very young embryonic tissue cells large star-shaped cells with various-formed processes; very few striated muscle fibres, very much modified. Microdactylia of both big toes, with aplasia of first phalanx.(?)

Case L.—Reported by Weldon Carter, in 1894. Male, aged five years. Family history neg. First symptoms appeared at 4 years of age—lumps in back. Never any complaint of pain or tenderness or inflammatory symptoms. Back and shoulders affected, especially latissimi dorsi. Both big toes in hallux valgus position; the phalanges of fourth and fifth toes were very short and slight webbing between second and third toes of each foot.

Case LI.—Reported by Carl Hochhalt, in 1894. Male. Family history neg. First symptoms appeared at 19 years of age—pains and swelling in right triceps brachii. Pains and swelling gradually disappeared after three months, leaving osseous hardening; similar attacks repeated annually, involving new groups of muscles. Spinal column is rigid and ankylotic in all its articulations. The muscles of the back are changed into hard bony plates, especially both cucullaris and latissimi dorsi, exostoses trunci; mimic muscles of face intact, masticating muscles rigid. In order to allow nutrition patient had several teeth broken out. Pushes nourishment through gap. Both shoulder-joints stiff, both triceps changed into hard bone masses. Under left gastrocnemius large bony mass size of one and a half fists reaching partly up into popliteal fossa, contracting knee-joint. Left glutæus changed entirely into shapeless bony mass. No congenital abnormalities mentioned.

Case LII.—Reported by E. Lexer, in 1895. Male, aged 50 years. Family history neg. First symptoms appeared at 35 years of age—pains in left side of chest. Pains disappeared after three weeks, leaving soft-covered nodules the size of hazelnuts at the painful places. These nodules also disappeared after a short time. In 1884 again pains in left side of chest and back. Soft swelling in left lumbar region size of palm of hand. Later grew smaller and harder. Till 1888 recurrence of symptoms every summer. The lumbar swelling grew larger and harder and spread in front and upward to the axillary line. He again fell sick in 1890, then remained free until 1894. New nodules had formed and a very severe recurrence took place, with pain in both shoulders and in muscles of both upper arms. In the lower part of the pectoralis major a cartilaginous nodulous tumor, size of a hen's egg. At the margin of the trapezius muscle, level of sixth cerv. a similar but smaller tumor. On the left deltoid a growth size of a man's fist, of fibrous consistency, covered by muscle and slightly movable over the underlying bone. Both upper arms show under atrophic biceps a cord-like, bone-hard mass, movable on humerus and occupying the place of the brachialis internus. The right forearm shows in its middle a bone-hard growth about 5 cm. long and wide, movable sideways under the supinator longus. Some of the other muscles show peculiar indurations. In the back the left latiss. dorsi nearly entirely pronated by a tumor formed by the amalgamation of a number of smaller ones, which reach from the crista ilii to the vertebral column and up to the scapula. In the region of the left infraspinatus a very large free tumor from spina scapulæ reaching to the acromion. In right serratus, over fifth and sixth ribs, a very hard flat tumor. No deformities of fingers or toes (specially mentioned). Excision of deltoid tumor. Ten days later the tumor over fifth and sixth ribs was excised. Patient left clinic December 6, 1894. Letter of March, 1895, mentions several muscles of neck and lower extremities involved. Microscopically the sections show acute and chronic inflammation and induration of the inter- and intra-muscular connective-tissue (perimysium) hypertroph. changes from which the formation of bone tissue starts. Atypical case. Commencement of disease very late. Ossification not shown.

Case LIII.—Reported by Stephen Paget, in 1895. Male, aged 7½ years. Family history negative. First symptom appeared at 4½ years of age—swelling behind left ear. Since that time various swellings and growths have appeared, some disappeared again, while others remained in different parts of his body. Holds head in wry-neck position from contraction of left sternocleido. A small nodule of bone in the anterior edge one inch above clavicle. In each pectoral muscle irregular bony nodules moving with the muscles. In the right pectoral they are dispersed all through the substance of the muscle and continuous with plates of bone under the deltoid and along the latiss. dorsi. Latiss. dorsi contains a sharp bony ridge in the axilla; the serratus magnus shows hard nodules at its origin. The bony masses in the l. pectoral. are also continuous with bone in the fascia beneath deltoid. Along each side of spinal column a hard bony ridge, more marked on left than right side, seemingly part of latiss. dorsi. From ant. fold of each axilla

a narrow hard cord, thickness of about No. 5 English catheter, runs right down the level of iliac crest; they are not bony. Both big toes are shortened and turned outward in metatarsal phalangeal joint, and the phalanges turn under the second toes. (Microdactylia-hallux valgus.)

Case LIV.—Reported by Fuerstner, in 1895. Female, aged 15 years. Family history neg. First symptoms appeared at eight years of age—nodules size of pigeon eggs appeared and disappeared again after five or six weeks. Since the age of eight or ten months hindrance in motions of neck and arms from nodules appearing in these regions. Position of head and vertebral column stiff; rigid walk. All movements of head very limited through tension of hardened but nowhere nodulated muscles of the neck. In the long muscles of the back, however, there are several hard bony thickenings. At the inner margin of the scapula there are several osseous protuberances and bony plates in the latiss. dorsi. The arms are flexed most of the time; cannot be abducted, as tendons of pectorales major are hard cords. Both biceps muscles are hard as well as presenting a peculiarly sharp tendon; the lower parts of triceps feel ossified. On the left side the same changes are present in a lesser degree. Abdominal muscles and lower extremities show but slight changes. During observation of this case a swelling occurred in the left biceps muscle within a few days. It was of a doughy consistency, the skin was slightly reddened and there was considerable pain upon pressure. A small piece of the biceps muscle was carefully excised and the *microscopical examination showed a very pronounced and intense hypertrophy of the interfibrillary connective tissue,* especially in the neighborhood of the blood-vessels, while the muscular tissue itself remained completely unchanged. Both thumbs and little fingers considerably smaller than normal. Microdactylia of both big toes.

Case LV (?).—Reported by Eichhorst, in 1895. Male, aged 24 years. Family history neg. First symptoms appeared during time of observation in hospital—swelling and hardening of muscles of left calf. Subsequent course of disease unknown. The swelling extended from the upper end of Achilles tendon 5 cm. into the muscles of the calf. Skin at first red and inflamed, afterwards resumed its natural state. While the affected parts of the muscles decreased somewhat in size they so increased in hardness that they felt like bone; the skin was movable over them, and in so doing crepitation was distinctly felt. Patient had spina bifida. No change of condition was felt when patient left hospital or was observed later; no progressive ossifications in other muscles. No deformity of fingers or toes. This, as well as the former case reported by Eichhorst's assistant (No. 37), is a doubtful, atypical case.

Case LVI.—Reported by O. Paget, in 1895, and W. P. Herringham, in 1899. Female, aged 5 years. Family history neg. Said to have suffered very intensely from rickets. First symptoms appeared three weeks before date of examination—lump in back, which gradually increased in size more rapidly since the last week. Subsequent course of disease unknown. Head fixed by two indurated nodules and hard sternocleido. Right pectoral, left pectoral and latiss. dorsi all in same condition. General health fair (pigeon-breasted); two growths, smooth, oval and firm, about 4 x 2″ symmetrically on each side of vertebral column just below inferior angle of scapula. Similar growth, size of large walnut, attached to inferior angle of each scapula. Ossified node on external edge of right bicipital groove. Ossified node on forehead, the latter due partly to inability of child to protect itself when falling forward. Bilateral microdactylia of big toes ascribed to absence of first phalanx. Observation in 1899 shows marked progress of disease on skull, abdominal walls and both ulnæ; movements of shoulder very restricted. Skiagram of big toes shows microdactylia and hallux valgus, due to: (1) Irregularity and shortness of metatarsal bones; (2) thickening of outer side of first phalanx, tilting bone outward; (3) synostosis of the first and unguinal phalanx at oblique angle. Exostoses on first phalanx of the right middle finger.

Case LVII.—Reported by Harry Lockwood, in 1896, and Raymond Crawford, in 1899. Male, aged 4½ years. Family history neg. First symptoms appeared at 2½ years of age—German measles shortly before malady showed itself. Subsequent course of disease unknown. Body and head bent forward. Masseters hardened; right and left pectorales major ossified and fixed. Large masses of hard material on both sides of spine Microdactylia of thumbs and toes.

Case LVIII.—Reported by Zoege v. Manteuffel, in 1896. Subsequent course of disease unknown. Demonstration of a skeleton of a case of myositis ossificans progressiva, where the products of the disease could be seen from head to foot. The osseous neoformations correspond to the course of the muscles. They had developed in their connective tissue nearly everywhere independently of the skeleton, so that they could not be ascribed an exostoses. A synostosis of the shortened phalanges of the big toes. The thickening of the mandibulæ seemed to be caused by an ossification of the muscular insertions. Microdactylia.

Case LIX.—Reported by Ludwig Pincus, in 1897. Male, aged 25 years. Family history neg. When 13 years old fell on back from a height of two metres. First symptoms appeared at 14 years of age—observed in nape of neck two little nodule-like glands, but they disappeared later spontaneously. Later back, shoulders and left arm again attacked after trauma. Causes of attacks are generally traumatic. After a prolonged quiet an attack of violent pains and swelling, first on the left, then on the right side. Disease particularly pronounced in neck, back and surroundings of neck, surroundings of axillary fossæ, masseters, upper arms and thighs. Scoliosis, caput obstipum, exostoses and hyperostoses. The right carpo-metacarpo articulatio pollicis ankylotic; thumb smaller than normal. Microdactylia of both big toes, with hallux valgus. Faradic contractibility diminished. Strong fibrillary spasmodic contractions spontaneously, but more pronounced when touched. Venous stasis, especially in legs.

Case LX.—Reported by Von Kryger, in 1898. Female, aged 4½ years. Family history negative. First symptoms appeared at 2½ years of age—with inflammatory symptoms, a painful bluish-red nodule formed on back a little to the left of twelfth dorsal vertebra. The first tumor decreased greatly and in its place a round osseous protuberance appeared. At various times this process, nearly always accompanied by inflammatory symptoms, spread over back, nape of neck and vertebral column, abdomen and hips. At last face and neck were involved. From the rigid vertebral column emanate long irregular bony ridges; shoulders drawn up, lower arms flexed at right angles, only slightly movable. Hands crossed across the abdomen; legs slightly flexed at hips; right one abducted, immovable. Head inclined toward front and left; face turned somewhat to right; muscles of the cheek and mouth are very hard and mouth can be opened only half an inch. Everywhere in muscles one finds tough cords or bony ridges, some of which are connected with the skeleton, others appear to lie free in the tissues and nearly always follow the course of the fibres, rarely crossing them. From the ribs, the vertebræ and the pelvis real exostoses project vertically to the skin. Microdactylia of both big toes. They appear to have only one phalanx, which stands in valgo position upon the metatarsus. (No X-ray picture.)

Case LXI.—Reported by Walter Stempel, in 1898. Female, aged 7 years (?). Family history neg. First symptoms appeared at three and a half years of age—swelling in sternocleidomastoidei; later in cucullaris and various other muscles of thorax and body. Swellings felt hard at first, got gradually softer and disappeared, as did many other hard swellings in various groups of muscles during the course of three and a half years of observation. Last examination, January, 1898, showed ossification of both cucullares at occiput. Ossification of scaleni. Spine quite stiff. Fixation of lower jaw through hardening of masseters. Microscopical examination of muscle in first stages of hard swelling showed a hypertrophy of the outer fibrillary tissue,

which by its rapid growth into the muscular tissue, separating and pulling the muscle fibres, causes an intramuscular hemorrhage, which, according to Stempel, represents the first cause of the swelling which is being followed by the fibrous tissue growth and later by ossification.

Case LXII.—Reported by A. Roth, in 1898. Female, age (?). Family history neg. First symptoms appeared between 1 year and 9 months and 2 years of age—swelling in left side of middle of back. Another swelling at the left side of the neck followed closely; both disappeared after inflammatory symptoms. A similar process developed on the right side of the back, followed by swellings of both arms from shoulders to elbows and of various other places, like chest, abdomen and thigh. After a recedence of the inflammatory symptoms the afflicted parts showed hard exostoses-like masses. Arms grew stiff and lately the face became swollen. Patient perfectly stiff, as if in tetanic rigidity. Two small exostoses, size of peas, above left ear. Maxillary joints ankylotic. Mouth can be opened only ½ cm. The entire cervical muscular group forms one hard mass. The upper arms are bound together by a hard bony ridge in the anterior and posterior height of the axillary folds, comprising latiss. dorsi and pectorales major. Both scapulæ are fixed. Superior and inferior scapularis of osseous hardness. In both upper and lower arm muscles various ossifications and enclosures of bone. Muscles of hands free, narrow strip of bone in musc. obliq. abdom. ext. On both sides, at origin of sartorius, ossification. Microdactylia of both big toes. Valgus position; only one phalanx reaching to first interphalangeal joint of second toe. Microscopical examination of a piece of new-formed osseous tissue shows the change of connective tissue to fibrous, cartilaginous and bony tissue. He believes in passive and active participation of periosteum, as he found hardly any bony pieces free in the muscular substance; also in a congenital diathesis brought into action by traumatism.

Case LXIII.—Reported by Mr. Jacoby, in 1898. Male, aged 27 years. Family history neg. First symptoms appeared at 19 years of age—pains in ankle-joints and legs. Four years later pains occurred in hips and neck, and only two and a half years ago induration and ossification in the neck, involving all the contour of the neck. Another ossified mass between pelvis and lower edge of arch of ribs. Slight hardness in masseters, but patient can still open mouth well. July 14th: diagnosis confirmed by X-ray pictures characterized it as a bone-muscle disease, which leads to the formation of exostoses and direct ossification of the muscular connective tissue. No mention of congenital abnormalities.

Case LXIV.—Reported by A. Salomoni, in 1898. Male, aged 12 years. Sound, healthy parents. First symptoms since birth—deformities in neck and arms. Gradual and steady progression. At the time of first observation, in 1897, deformities had greatly progressed. They occupied the entire neck and the two upper limbs in a very characteristic manner. A personal letter from Piacenza, Italy, where Salomoni is now head of a military hospital, explains that on account of being so taken up with his present duties he is unable to go further into the history of this case, and merely gives me what data he remembers. He says: "I extirpated from the neck and from the arm two pieces of the muscles where ossification had gone on. They were histologically demonstrated to be derived from the fibro-connective tissue and from the new-formed bone near the periosteum. The little patient underwent various pharmaceutical treatments, more for the moral effect than for any other purpose, and died at the age of 14 without my being able to continue the observation of his case." This case was reported in the record of the congress of the Italian Surgical Society of Turin, 1898. Salomoni's letter differs from the above-named report and that of the Italian Pathological Society in 1898, which reads: "A girl, 7 years; when 2 years old ossif. of l. sternocleido noticed; later r. side; then gradual swelling and ossif. of other muscles, sometimes with fever. Microdact. of thumbs and big toes."

Case LXV.—Reported by Hendrick Burgerhout, in 1898. Male, aged 40 years. Family history neg. First symptoms appeared at birth—left shoulder-blade and left shoulder-joint stiff. During schooling years hip- and knee-joints became stiff. Later, after twentieth year, both elbow and wrist-joints stiffened. Ten years ago both maxillary articulations became stiffened so that he could not open mouth. Sits nearly immovable; head bent forward upon chest. Lower jaws very slightly movable. Distinct movement of temporales and masseters. Sternocleido and cucullaris feel hardened, but not osseous. Arms stand in strong adduction; scapulæ fixed; at lower outer angle not separable from the stone-hard mass which goes in the direction of teres majores and latiss. dorsi. The long muscles of the neck are connected from neck to os sacrum by irregular ridges of bone. Left arm, shoulder and elbow stiffened; left thigh flexed in hip-joint; left knee-joint and ankle-joint immovable. Large exostoses above patella in location of vast. externus. Right leg articulations resemble left side, but are freer. On inside of femur in place of adduct. hard exostoses extending from pelvis to insertion of adductor magnus. Microdactylia of thumbs and big toes due to smallness of metacarpal and metatarsal bones. Very exact examination of electric irritability of muscles and their nerves; both normal. For very thorough metabolical research work and conclusions of this case see text.

Case LXVI.—Reported by Robert Jones, in 1899. Male, aged 16 years. Family history neg. Fell and injured his back.(?) First symptoms appeared at 13 years of age—growths on back started one year after his fall. Subsequent course of disease unknown. Growths on inner aspect of right jaw; small lump below right olecranon. Hard mass attached to upper two-thirds of femur; thickening at insertion of right ligamentum patellæ and at each tarsometatarsal joint. Over spinous processes a continuous line of hard cartilaginous or osseous material, extending from level of fifth dorsal to fourth lumbar spine. Strips projected from the upper and lower ends of this mass two and three inches long. They were movable in the surrounding tissues. No deformities mentioned.

Case LXVII.—Reported by Morian, in 1899. Male, aged 4½ years. Family history neg. Fell twice down a flight of fourteen steps. First symptoms appeared at 2½ years of age—soon after second fall swellings in various muscular groups. Fell again after commencement of disease and struck on a stone with his neck. Ossification in right frontalis muscle, in both sternocleid., in the group of muscles from sternum to hyoid and from there to mandibula, in both pectorales major and deltoids, in both sacrolumbales and in both bicipites; in both cucullares and right altissimi dorsi. Both big toes show microdactylia.

Case LXVIII.—Reported by Lyder Nicolaysen, in 1899. Female, aged 4½ years. Family history neg. First symptoms appeared at 2 years of age—stiffness of shoulder, neck and back. Subsequent course of disease unknown. Height 99 cm. Circumference of chest 45 cm. Posture: head bent forward; nape of neck, back and shoulder muscles completely ossified. In both axillary fossæ anterior and posterior folds are hard; greater part of pectoral. maj. ossified. Both scapulæ fixed to thorax. Trapezius bone-hard. Several nodular exostoses on back. Arms stiff and rigid. Hardened parts in musculature of upper arms. Excursion angles of both forearms about 45 degrees. Microdactylia of both thumbs and big toes.

Case LXIX.—Reported by George Wilkinson, in 1900. Male, aged 13 years. Family history neg. First symptoms appeared at 2½ years—swelling in right side of back. Swelling in back subsided. Later isolated hard lumps appeared on back, remaining and spreading. Later stiffness in back. Lower jaw fixed, mouth can only be opened one-fourth inch, due to hard temporal fascia. Bony bars in right occipital bone. Thin plate of bone in occipital tendons and left trapezius, and hardening in right. Hard nodules in both sternocleidii and scaleni muscles. Big bony mass extending from left to right scapula. Two large hard masses in right lat., shoulders raised, scapulæ fixed. Nodule in left pectoral. maj.; two small ones on both

sixth ribs. Respiration wholly diaphragmatic; shoulder movements, especially rotation, limited. On each femur large irregular mass of bone fixed to the upper end near great trochanter. A spike of bone extends on each side of glutæus maximus near its sacral origin. Another one down along outer side in tendon of fascia lata to below middle of thigh. Hard nodule in right rectus femor. Sharp nodule of bone projects below each tuberosity ishii. Angular bony nodule on posterior border of right tibia, three inches above int. malleolus. Movements of hips limited. Trunk bent somewhat forward, can sit only on corner of chair. Both thumbs microdactylic. Their metacarpals and phalanges are shorter than those of other digits and interphalangeal joints stiff. Both big toes microdactylic and with hallux valgus. Their two phalanges very short, their interphalangeal joints synostotic; nodules have formed without pain. Some masses that have appeared during the nine months of observation have disappeared again.

Case LXX.—Reported by Morley Fletcher, in 1900. Male, aged 9 years. Family history neg. First symptoms appeared at 7 years of age—location (?). Subsequent course of disease unknown. Numerous exostoses on bones of head, one on the hand and one on each tibia. Thickening of bridge of nose. Both big toes small and terminal phalangeal joints ankylosed. Interesting association of numerous exostoses with multiple neuro-fibromata; looks upon it as abnormal condition of mesoblast from which the mesoblastic structures were formed and as related to the condition of multiple exostoses and multiple neuro-fibromata.

Case LXXI.—Reported by Theodore Schwickerath, in 1901. Female, aged 14 years. Family history neg. First symptoms appeared at 9 years of age—violent toothache. Two molars were extracted, whole left side of face swelled greatly after extraction of teeth and became permanently stiffened. Patient's jaws became so stiff that she could not open her mouth sufficiently for mastication. A piece of an osseous ridge 1 cm. broad and ½ cm. thick was removed from the left masseter to increase the possible excursion of the lower jaw. After the operation mouth could be opened 3 cm., but later it returned to almost its preoperative status. Patient holds herself bent forward. Scar on left masseter where excision was made. Near upper insertion of left masseter a firm nodule, size of cherry pit; in its lower part an osseous ridge 1½ to 2 cm. broad. Lower jaw can only be opened 2 mm. Lateral movements entirely inhibited. Left clavicle enormously thickened. Left sternocleido a thick cord, shows osseous ridges in sternal and clavicular portion of the pectoral. maj., is as hard as bone, running toward axilla and ending at spine of tuberosity maj. In the middle of right scapula spine is an osseous nodule size of a walnut. Back scoliotic to left and shows partly flat, partly osseous protuberances. On both sides of vertebral column osseous plates; in latiss. dorsi and teres osseous nodules extending right down to region of sacrolumbalis. Right arm: very few excursions possible; adduction, abduction and rotation impossible; in the axilla a hard nodule. Left arm nearly normal; right thigh exostotic thickenings. Microdactylia of both big toes. No X-ray picture.

Case LXXII.—Reported by Vaughan and Burton Flanning, in 1901. Male, aged 33 years. Father had same disease, dating from a fall in his fifteenth year on his right arm, where it began. The disease had steadily progressed, but had not reached the same degree as with his parent, when he died at the age of 33 from an accident. He had congenital deformity of both thumbs, there being no joint beyond metacarpal ones. Feet were not observed. First symptoms appeared at 8 years of age—left shoulder gradually became stiff without any previous injury or pain. At 12 years right knee began to stiffen and gradually became ankylosed. Following this came stiffness of left hip, sides of chest and lower part of back. At 26 years right arm from shoulder downward became swollen, red and tender. This disappeared, but left arm is fixed extended in shoulder and elbow. Wrist remained free. Two years later stiffening of right hip was noticed. During winter of 1898 difficulty in

opening mouth began; passed off during summer, but returned worse the following winter. The following summer got slightly better, but had gradually increased since. Both erector spinæ nearly completely replaced by bone, preventing any movement of that portion of the spine covered by them. Deep muscles of back of neck similarly affected, only possible movement of head is slight rotation and flexion. Bony plates between several ribs; on each side and along ant. border of l. quadrat. lumb. was hard band fixing twelfth rib to crest of ilium. Movement of chest almost prevented by these growths except over first two ribs. The distal halves of both teres maj. and pect. maj. were bony. Close over the right fossa olecrani a bony mass, the size of a walnut, involved the whole thickness of the triceps. A similar growth over the upper ends of the ulnæ and radii. Both extens. quadriceps were almost replaced by bone; osseous growths round head of right fibula and enlargement of both int. malleoli. Both big toes had hallux valgus, with shortening and broadening of phalanges, which were fused together. No X-ray.

Case LXXIII.—Reported by Wilhelm Rager, in 1901. Female, aged 14 years. Family history neg. First symptoms appeared at 2 years of age—swelling over os frontal. After that many painful inflammatory swellings in various parts of the body, leaving osseous hardenings. Mouth can only be opened 1½ cm. Both shoulder-joints and right elbow-joint ankylotic. Ossification in both masseters, right sterno-cleido and platysma, both deltoids, pectoral. maj. and latiss. dorsi, quadriceps femoris, right soleus and insertions of both gastrocnemii and solei. Similar ossifications are distributed all over back of neck and the back proper down to lumbar vertebræ. Both thumbs and big toes are deformed, only half as long as normal. In the thumbs, due to two shortened and synostotically united phalanges; in the toes to stunted metatarsal bones.

Case LXXIV.—Reported by Christopher Graham, in 1901. Female, aged 6 years. Family history neg. First symptoms appeared at 4 years of age—small lump between shoulders. Gradual but steady progression. Rarely any pain except during last three or four months. Ligamentum nuchæ solid throughout. Both pectorales and sternocleidi are more or less solidly ossified. Right biceps affected in its entire course; the left only in its upper part. Right trapezius is completely changed into bony mass; the left less so. Both latiss. dorsi are ossified. The deeper trunk muscles seem to be also similarly affected. Right external oblique ossified along the line of the linea semilunaris, below and externally from Poupart's ligament, a hardness and nodules can be felt. No microdactylia mentioned. Upon my inquiry, Doctor Graham kindly answered that he had no recollection of having examined the feet, the boy having been seen in the policlinical department only.

Case LXXV.—Reported by H. D. Rolleston, in 1901. Male, aged 8 years. Subsequent course of disease unknown. Ossification in both sternomastoids and posterior scaleni muscles and in the scapular attachments of the rhomboids. Bony plates in lower parts of latiss. dorsi. Hard masses in region of erectores spinæ over lumbar spine. Both biceps and brachialis ant. muscles extensively ossified. Both recti abd. are affected. Exostoses on both tibiæ near insertion of sartorius muscle and on both frontal bones. Microdactylia of both thumbs and toes, due to dwarfing of metacarpal bones. Commencing ankyloses of interphalangeal joint. Both hallux valgus, with exostoses. Exostoses on first phalanx of right middle finger. (Same peculiarity in Doctor Herringham's case, No. 56.)

Case LXXVI.—Reported by George Carpenter and Walter Edmunds, in 1901, and Cyril Nitch, in 1908. Female, aged 4 years. Family history neg. First symptoms appeared at 3½ years of age—two weeks after both tonsils were removed a hard and tender swelling appeared on right side of neck. The first swelling subsided after a couple of weeks. Two weeks later a swelling appeared on left side, which also disappeared again. A short time afterward the mother noticed a hard mass on right side of neck, and a few months later similar bumps in left pectoral,

scapular and lumbar region. *Condition in 1901:* Spicule of bone in left sternocleido. The neck muscles of same side infiltrated and hard. Below chin bony infiltration size of pea in geniohyoid muscle. Plates of bone can be felt in right sternohyoid, right coracobrachialis, left pectoralis major and erectores spinæ. February, 1905: back and neck quite rigid; movements of shoulders very limited. The early pea-like prominence of bone in geniohyoid has now become a long spinous process. Osteoid tissue in form of plaques, bosses and spicules can be felt in both erectores spinæ, latiss. dorsi, trapezii and pectorales, right rhomboideus major and minor, the sternomastoids, right vastus externus and in popliteal space. Microdactylia of both big toes. 1901: excision from a muscle that showed gray infiltration revealed under microscope fibrocellular tissue infiltration; no micro-organisms.

Case LXXVII.—Reported by Menard and Tillaye, in 1902, and Comby and Davél, in 1904. Female, aged 5½ years. Family history neg. First symptoms appeared at 17 months of age—swelling (where?). Swelling always preceded hardenings and ossifications (where?). Osseous hardenings in post. cerv. regions and down the back to right axilla, extending toward thorax and to the inner border of humerus and also to the dorsalis maj. On the left side similar changes in the trapezius. Two and a half years later: slow, but steady development of new osseous hardenings. Stalactite formations in axilla, reaching down along humerus to elbow. Head, arms and back stiff, and movements extremely limited. The thumbs and little fingers of both hands as well as the two big toes are undersized. (Microdactylia.) Woodcuts of hands in later observation show microdactylia of thumbs depends on shortness of metacarpal bones. No X-ray in either communication.

Case LXXVIII.—Reported by Fabio Rialta, in 1902. Female, aged 16 years. Family history neg. Father suffered for thirty years from arthritis of the left hand. Vaccination (?). First symptoms appeared at 2 years. After vaccination swellings appeared on head, from size of pigeon's to hen's egg. The entire left arm became swollen, then subsided. The same occurred then on right arm. Later both arms contracted, with flexion of forearm and adduction of upper arm and inability to raise them. A gradual, continual increase of all these symptoms. The neck swelled and remained stiff. After swelling had subsided some hard indolent nodules developed over the dorsal spine of the scapula the size of those on the head. From the superior dorsal region these formations descended, increasing in number every spring for about twelve years until two years ago, when they stopped. Height 138 cm. Head flexed rigidly toward right and forward; neither active nor passive rotation, extension or flexion of head possible. Vertebral column equally rigid; muscles of neck atrophic; sternocleido contracted and hard. The posterior cervical ligament ossified, with osseous plate; another one runs from seventh cervical downward; ossification of left levator scapulæ and cucullaris. The tendon of the pectoralis major is ossified to its insertion. Arms crossed over chest; forearms in exaggerated flexion; osseous plates in biceps, the caput brevis completely enveloped with an osseous growth the size of palm of hand. The anterior wall of axilla also ossified and posteriorly is an ossification extending on both sides from dorsal. maj. to scapular angle and to vertebral column. R. biceps has ossification similar to but less pronounced than left. Processi spin. below seventh dorsal are hidden by ossified masses. Column is slightly scoliotic and absolutely rigid. Respiration completely abdominal, slightly thoracic at base. External genitals normal in size, but hairless. Microdactylia of both big toes. Shortness of first metatarsal bone and hallux valgus. Hemidrosis facialis (sinistra).

Case LXXIX.—Reported by M. Ferraton, in 1903. Male, aged 18 years. Family history neg. First symptoms appeared at 3 years of age—previous to first swelling in scapular region a three months' period of pain, sleeplessness and loss of appetite. After swelling subsided ossification set in. Ossification of right masseter; left more atrophied. Right sternocleido hard; the greater part seems to be ossified. Pect

maj. and deltoids contracted and atrophied. On the right humerus anteriorly two round exostoses, size of hazelnut, firm upon the bone; on the left side similar outgrowth. Another exostosis on the pelvis on crista iliac. post. to its edge, close to ossified insertion of dorsalis maj. Several large ossifications on the inner side of the right thigh, under middle and large adductors. Right foot: second toe much larger than the others, microdactylia of big toe; on left foot a synovial bursa on first phalanx of big toe. Hallux valgus. (No X-ray.) Operation on masseter muscle; exsection of small piece of muscle. Patient can open his mouth.

Case LXXX.—Reported by Julius Michelson, in 1904. Female, aged 18 years. Family history neg. First symptoms appeared at 7 years of age—both thumbs over both flexor. brev. hot and inflamed. Later thumbs grew stiff and nearly immovable. Kyphosis and scoliosis at cervical and dorsal spine. Cervical spine stiff, only minimal movements possible in rotation, extension and flexion. Same conditions prevail in dorsal and lumbar portions. Muscles of neck hard. (X-ray shows no ossification.) Pars clavicularis of left pectoralis maj. shows bony deposit extending over the shoulder-joint and into the axilla; this mass can be moved to and fro. A similar mass is on the right side. Both thumbs are ankylotic between first phalanx and metacarpus. The little fingers are shortened. (Second and third phalanges are shortened.) Both big toes are 2 cm. shorter than normal and their two phalanges in firm osseous union. The little toes also are shortened and their second and third phalanges ankylotic.

Case LXXXI.—Reported by Clito Salvetti, in 1904. Female, aged 4½ years. Family history neg. First symptoms appeared at 10 to 12 months of age—swelling observed in the left cervical region. This swelling disappeared, but was followed by two elastic swellings on the occiput, which persisted until now. Neck stiff. Hard infiltrations of muscles back of both supra- and infra-spinosi, and tumor-like indurations size of hen's eggs. Left pectorals like an inflexible plate, preventing the shoulder movements entirely and those of the forearm partially. The left Mohrenheim fossa absent. Neither fingers nor toes show microdactylia. (Specially mentioned.)

Case LXXXII.—Reported by A. P. Beddard, in 1905. Male, aged 37 years. Family history neg. First symptoms appeared at 7 years of age—bony growths began to develop in muscles. Subsequent course of disease unknown. Bony growths can be felt in innumerable parts of his muscular system. Some of them are movable within the muscles; the majority are fixed, having developed in tendons and insertions of muscles. Ankylosis of joints has developed to such an extent that patient is entirely helpless and absolutely bed-ridden. No mention of microdactylia.

Case LXXXIII.—Reported by A. Gaster, in 1905. Male, age (?). Grandfather. father and three sons were afflicted with same disease. Subsequent course of disease unknown. Doctor Gaster states in the discussion of Doctor Beddard's case (presented by Doctor Russel) that he knows a family in which the father and grandfather had myositis ossificans progressiva. The three sons suffer from the same disease. Mother and two daughters are free from it; two sons have two baby daughters without a trace of the disease.

Case LXXXIV.—Reported by Nové Josserand and René Horand, in 1905 and 1912. Female, aged 7½ years. Family history neg. First symptoms appeared at about 6½ years of age. Subsequent course of disease unknown. On both sides of scapulæ a tumor size of mandarin, the right a little larger than the left. Osseous indurations on the trapezius and in the upper part of right sternocleido. In lumbar region an osseous plaque near the iliac crest. In right and left pectorales major and minor are ossifications. Both big toes are shortened and show hallux valgus. Last phalanx of each little finger is noticeably turned outward. In 1912, on account of progression of disease, patient was induced to submit to X-ray treatments. Had twenty-six treatments, all below five Holzknecht's units. Effect surprising; patient straightened out,

can again walk with youthful agility up and down stairs, sit and lie down unaided, and is no more imprisoned in her shell. As to duration of this improvement, author promises watchful waiting.

Case LXXXV.—Reported by S. Biegel, in 1906. Male, aged 7 years. Family history neg. Born in occipital presentation; was delivered with forceps; large hæmatoma over occiput (?) Head at first drawn out lengthwise, but after few days normal shape restored. First symptoms appeared at 3 months of age—parents observed apparent displacement of parietal and other bones of skull, and a little later swellings on head size of pea to a marble. These swellings disappeared after a few weeks. Previous to appearance of swellings on top of head and on neck there was always stiffness vertebral column and cyanosis of head and of extremities, which again disappeared as soon as swellings were fully developed. This lasted with intervals to his third year. From third to sixth year he was quite well, although a slight knock or a slight fall always produced hard nodules, which disappeared again after a few days. About one and a half years ago he fell upon his head. From this time on he grew stiff. Can now hardly move. At first his neck grew stiff and he could not turn his head. Later maxillary muscles stiffened; mouth could not be opened more than enough to admit flat spoon. This condition improved so that he could open his mouth again, but stiffness developed in the muscles of his mouth. *September, 1905:* a stone-hard thickening of both sternocleidei; *December*, stiffening of upper arm and thorax muscles of both sides, in left side osseous ridge remained. Head bent to left side and forward; breast and abdomen bent backward and to the right. Both sternocleidei and platysmæ feel like hard plaques. From left side of axilla a bony ridge goes over pectoralis major, serratus ant. maj., obliquus ext. abd. Both cucullaris are hard, the edge of the scapulæ strongly projecting. The right arm hangs along the side of the trunk and the hand rests on the thigh; the left is abducted in shoulder-joint and flexed in elbow so that the hand rests nearer to Poupart's ligament. Both are limited in their movements, the left more than the right. All the hand and finger-joints are well movable. Both legs are flexed in hip- and knee-joints. Hip-joints are stiff, but below these joints limbs are well movable. Microdactylia of both thumbs and big toes.

Case LXXXVI.—Reported by E. J. Maxwell, in 1907. Male, aged 10 years. Family history neg. First symptoms appeared at 5 years of age—hardness of spine muscles. Gradually increasing ossification of muscles of spine near the sacrum and back, spreading upward. Spine and head absolutely rigid; growths of bones branched out from spine to scapula. Breathing mainly abdominal. Only liquid could be swallowed; condition of masseters and jaws not given. Bone could be felt on right side of larynx. No mention of microdactylia. No X-ray picture.

Case LXXXVII.—Reported by A. E. Garrod, in 1907. Male, aged 21 months (weight 23½ pounds). Family history neg. First symptoms appeared at 3 or 4 months of age—lumps appeared on head. Child healthy at birth, but soon began to waste. Improved to normal condition, however, when nursed by colored woman. The lumps disappeared completely three or four months later. At 16 months fresh swellings appeared upon back, increasing in size for some time, then disappeared again, while fresh swellings had appeared in new locations. Swellings upon back, two in front of chest in outer parts of pectoral muscles; one on the head over occipital bone. Swellings disappeared and new ones came during the time of observation, *May 29, 1906, to July 20, 1906* Patient left without any bone formation having occurred in any of the swellings during this first period of observation; no constitutional disturbance or particular pain seemed to accompany the appearance of the swellings. One year later reported: right biceps is contracted and feels stony hard; stiff elbow and right shoulder-joint. A lump size of mandarin orange over infraspinatus, of ivory hardness. Other lumps forming. Health seems to be perfect. Microdactylia of both big toes in valgus position. Over the inner aspect of the head of

each first metatarsal bone was a small linear scar, suggesting that an extra digit had been removed.

Case LXXXVIII.—Reported by Paul Krause and Max Trappe, in 1907. Female, aged 16½ years. Family history neg. Severe cold (?). First symptoms appeared at 12½ years of age—tired feeling in arms and legs, and back began to feel stiff. Pains in various parts of her body; knees sensitive to pressure; stiffness of legs increased, arms became bent and stiff. About the winter of 1905-06 appeared a swelling in lower sacral region, which disturbed the patient when lying down. A bone-hard nodule was removed from right ant. axillary wall; since then she can move right arm better than left. A swelling of the feet appears now from time to time, but soon disappears again. The swellings when softened evacuated an emulsion of amorphous calcium salts. Later the skin became sclerotic, dry and immovable over its underlying tissues. Patient 155 cm. tall; weight 34.5 kil. Very strongly developed arci zygomatici and large pupillary distance (65 mm.). Slight left scoliosis in lower dorsal and lumbar portion. Pect. tendons are hard as board, contain several nodules hard as cartilage. Scapulæ show over spinæ several small tough nodules; over right acromion a hard tumor size of a cherry pit. In upper and lower parts of both biceps are several small nodular imbeddings. In right cubital fossa a collection of nodules connected with the lacert. fibrosus, continuing to the forearm; on the left only one small nodule. *Hands show nothing abnormal.* Over right sacro-iliac joint nodule of bony (?) consistency, size of millet grain, and to the left of the spinous process of fourth and fifth lumb. vert. a soft swelling, size of a silver dollar, sensitive to touch. On both sides of os coccyx three globular swellings about 2½ cm. In the gluteal muscles small nodules size of cherry pits. The adductors at their origin, at os pubis, as hard as board. At the inner epicondylus of the left thigh two nodules of cherry-pit size adherent to skin and movable with it over the underlying tissue. Toes all well developed. The microscopical examination of the excised axillary nodule showed only a sort of fibrous hyperplasia and infiltration of the muscular tissue. The late beginning (12½ years), after a cold, the absence of new pathological cartilage- or bone-formation or even of calcification in the X-ray pictures or in the excised nodule, the softening, liquefaction and discharge of thick, mucous emulsions, the dryness, sclerosis and immovability of the skin, the absence of micro-dactylia—all these features do not fit into the picture of myositis ossificans progressiva, but stamp the case as belonging to *myositis fibrosa.* Since writing I found that in a later article the authors* abandoned a diagnosis of this case as myositis ossif. progressiva.

Case LXXXIX.—Reported by Roberto Solè, in 1908. Female, aged 7 years. Family history neg. First symptoms appeared at 4 years of age—parents detected two cords in abdominal wall extending from thorax to pubes, and according to description of location were the recti muscles. They felt hard to the touch. These two cords persisted in their hardness till child was 4 years old, when they gradually disappeared and abdomen grew normal. Often fell in the first years of her life and at 2½ years had severe fall, after which and since numerous hard tumors appeared, mostly on neck and thorax. Suffered severe pains at 3 years. These disappeared at 4 years, but tumors remained and increased in number and size. Head bent forward and fixed firmly. In the ant. region of the neck two stiff hard cords in the location and direction of the sternohyoids and mylohyoids, like rosaries on account of their interspersed osseous infiltrations along their course. A similar formation exists porteriorly on both sides of the cervical vertebral column, from the superior part of the thorax to the base of the occiput. Thorax in kyphosis position, but not angular. It shows numerous hard osseous tumors over the ribs, some intimately adherent to them; others simply on their borders. Both pectorales major and latiss. dorsi are ossified, keeping the arms in their rigid posture in the shoulder-joints. Some

* Fortschr. auf d. Geb. d. Röntgen Strahlen, bd. 14, H. 3.

few nodules of ossified tissue in triceps brachii. No mention of microdactylia, but in one of the photos of this case the presence of microdactylia of the big toes can be plainly seen.

Case XC.—Reported by Warren Walker, in 1908. Female, aged 7 years. Family history neg. First symptom appeared at 6 years of age—stiffness of neck. Later a swelling appeared in the right pectoral muscle and others in different parts of the body. These swellings were painful to the touch. Marked prominence of occipital tendon of right trapezius, muscle very hard and stands out like a cord. The scaleni and sternocleidi also involved, but not so extensively. Head fixed on sternum, no rotation possible. At lower angle of left scapula exostosis one inch high, another one over ninth rib just below the scapula is covered by a mass of indurated muscles. Right scapula similar to left. Exostoses on eleventh and twelfth left ribs and several on left of lumbar spine in substance of erector spinæ. Spinal column rigid. Arms are held off trunk in abduction, cannot be brought to side of trunk. Left humerus only movable with aid of scapula, over shaft a bony mass size of pigeon egg. Motion of right arm also limited. Bony masses in anterior and post. axillary folds prevent motion. Entire absence of thoracic respiratory movement. Breathing abdominal muscles free. Left thigh 1.75 cc. larger than right. Fingers are all shorter than normal; little finger particularly so. Hallux valgus and microdactylia of both big toes. The first metatarsal bones short and stubby and interphalangeal joints of big toes ankylosed.

Case XCI.—Reported by Charles Adair Dighton, in 1908. Male, aged 11 years. Family history neg. First symptoms appeared between 4 and 5 years of age—following successive attacks of measles, bronchitis and whooping cough; complained of general malaise and weakness—most marked in legs; child unable to walk. Subsequently muscles became stiff and hard, lumps formed in them, increasing steadily in size and numbers up to time of observation. Weak and undergrown. Skin very tender to touch. Walks on balls of toes. Lost almost all movements of shoulders and elbows. Both arms show large bony nodules in biceps, triceps, coracobrachialis and anconæus. The biceps are almost completely converted into bone. Left arm shows greater development of osseous masses. Forearm free, but muscles atrophied. In both legs adductors completely ossified. Extensors and flexors less affected. Left leg more marked than right. In the line of fibres of obliquus abd. ext. on either side where they run toward the pubic bone is an osseous plate the size of a man's hand. No mention made of microdactylia. Doctor Dighton kindly sent me his notes of the case, but no mention was made therein of presence or absence of microdactylia.

Case XCII.—Reported by Charles F. Painter and John D. Clarke. Male, aged 25 years. Family history neg. First symptom appeared at 6 years of age—torticollis; a symptom pronounced as hereditary in his family. Numerous nodules developed all over the body in various parts and different articulations of spine and limbs became ankylosed. The right hip was mobilized operatively by removal of bone so that the joint could be freely moved after operation, but bony masses soon formed again. An open incision tenotomy of internal and external hamstrings was equally unsuccessful. A hard mass on right side of chest. A mass the size of a lemon situated under the left glenoid cavity. Motion of elbow limited. Numerous nodes can be felt over arms, wrists and fingers near joints. On both sides of the spine over the fourth or fifth ribs are pronounced nodes, and similar ones occupy the right side of spine opposite sixth to eighth ribs. Slight motion in atlanto-axoidean articulation, restricted by nodes. No thoracic respiratory motion. Dorsal and lumbar spine completely ankylosed. Left hip flexed about 40 degrees and adducted, with practically no motion. Nodes on left tibia on the upper epiphysis, limiting both extension and flexion of knee. Left ankle ankylosed, with nodes on dorsum and planta of metatarsus and phalanges. The right quadriceps muscle and the hamstring tendons

undergoing ossification along their entire length. Nodes on tibia similar to left side. Right foot similar to left. The second toe of the left and second and third toes of the right foot are mentioned as being enlarged. (The X-ray picture of the left foot shows microdactylia of big toe through smallness of metatarsal bone.) This and a similar condition in the right foot may explain the elongation (?) of second and third toes.

Case XCIII.—Reported by G. Rizzuto. Female, aged 8 years. First symptoms appeared at 4 years of age—in greater part of muscles of neck and nape of neck. Later attacked sacrolumbar regions. Osseous hardenings in neck, lumbar region, scapula, thorax and arm. Later affected the roots of right last ribs over the corresponding iliopsoas. Made hæmatological observations. (Only this brief, incomplete history as an account of the demonstration in the official report of the congress.)

Case XCIV.—Reported by H. B. Allen, in 1909. Male, aged 27 years. Family history neg. First symptoms appeared in early youth (?). Subsequent course of disease unknown. Right temporal muscle partly ossified. Plates of bone along main tendon of erector spinæ in dorsolumbar region and spreading in direction of supraspinous ligaments, serrati post. inferior, and from lower ribs to inferior angle of scapulæ, uniting them both and sending off superficial process into ossified ligamentum nuchæ. Left arm: thickening of bone at inner bicipital ridge, at posterior edge of ulna and around wrist. Right arm: thick buttress of bone in pectoralis major from clavicle to outer bicipital ridge. Beginning ossification in capsule of shoulder. Osseous plates along shaft of humerus and process from upper part of olecranon. Osseous hypertrophies at upper and lower ends of forearm. Left leg: thick buttress bone from ischium to femur, in line with quadrat. fem. Some osseous hypertrophies near insertion of glutæus maximus and upper part of adductor longus; osseous growth with knee-joint, causing ankylosis, also binding the heads of tibia and fibula; similar growth at lower ends of bones. Right leg: bony ankylosis of hip-joint, with outgrowths from trochanter reaching tuberos. ischii, and others from both trochanters to front of os pubis. Lower ends of bone similar condition as left. Only photographs and skeleton available. Microdactylia of both thumbs and big toes and ankylosis between some carpal and interphalangeal bones; ankylosed and deformed second and third toe-phalanges.

Case XCV.—Reported by L. R. Krever, in 1910. Female, aged 28 years. Family history neg. First symptoms appeared in twentieth year—very painful swellings on left shoulder, with light chilblains. Subsequent course of disease unknown. Ossification of pectorales, deltoid, supra- and infra-spinati muscles, trapezii, latiss. dorsi, the lumbar muscles, fascia lumbodorsalis. No microdactylia mentioned.

Case XCVI.—Reported by K. F. Person, in 1910. Male, aged 18 years. Family history neg. First symptoms appeared at 5 years of age—soft, painful tumors on sides of neck. Tumors disappeared later, only to reappear in back of neck. At 13 years of age ossification of posterior muscles of neck, strongly involving ligam. nuchæ, spinal and lumbar muscles, and erectores of left thigh. Head slightly inclined backward and completely immovable. Spinal column has slight scoliosis, is completely immovable in cervical and dorsal portions. Slightly movable in lumbar portion. Exostoses and hyperostoses on scapula, vertebræ and head of left fibula; sharp atrophy of muscles of back, scapula, pelvis, which have not become ossified. Limited movements of both shoulder-joints, especially right; of both knees, especially left; of both ankle-joints and of toes. Scapulæ and ribs completely immovable; respiration abdominal. Microdactylia of both big toes, with aplasia of phalanges; bilateral hallux valgus and bilateral partial syndactylism of second and third toes.

Case XCVII.—Reported by G. A. Pirie, in 1910. Male, aged 6 years. Family history neg. First symptoms appeared at 2 years of age—could not turn his head freely; had difficulty in moving it upward. Thickening and induration of ligamentum

nuchæ. Hardness gradually increased up and downward, and when child was 3 years old the new tissue felt as hard as bone and had extended and adhered to occiput. This osseous growth was removed, and thereby the absence of spines and laminæ of third and fourth cervical vertebræ and defect in the closure of the spinal canal revealed; the spinal cord was exposed. A few months after operation difficulty in raising his arms developed, and examination showed swellings and indurations in latiss. dorsi muscles along both sides of vertebral column. They gradually increased and spread until nearly all the muscles, tendons and ligamentous tissues over back of thorax have become ossified. Osseous formation on back of neck has been formed afresh and is adherent to vertebræ lower down. A similar one is forming on left side. On right side osseous band extends from lower rib to crest of ilium. A few bony nodules at inner sides of both tibiæ and slight induration of adjacent muscles. Microdactylia of both thumbs.

Case XCVIII.—Reported by Alves de Faria, in 1910, and Jorge de Toledo Dodsworth, in 1912. Sex unknown. First symptoms and age of their appearance unknown. Subsequent course of disease unknown. Entire report based on two radiographs. 1. Taken in left profile: osseous changes in superficial muscles of neck, after severe atrophic changes. Osseous bridge from occipital protuberance to dorsal muscles. Between these and vertebral column some indefinite shadows, denoting second stage of invasion. 2. Anterior view of thorax: from seventh left intercostal space running toward humerus, but not reaching it, was a cross-formed osseous process. On right side a similar bridge joins the bone. From sixth rib a bony elongation runs to the outside and ends about middle of preceding one. The two are connected by a third one. Humerus thickened in upper part; numerous osseous nodules on back. Microdactylia not mentioned. No X-ray picture of hands or feet.

Case XCIX.—Reported by P. J. Stoyanoff, in 1912. Male, aged 25 years. Family history neg. First symptoms appeared at 10 years of age—when patient had variola. Subsequent course of disease unknown. Signs of former variola all over body. Insertion genioglossus muscles ossified. Neck rigid and moves with great difficulty. Trapezii ossified and joined to scapulæ, which stands out wing-like. Scaleni ossified. The clavicles are bent, but muscles of the supra- and infra-spinous fossæ being ossified. Multiple exostoses; axillæ ossified. Large portion of pectorales ossified. Insertion of deltoids ossified, as also that of left triceps humeri. Parts of biceps and coracobrachialis ossified. Left elbow ankylosed at 180 degrees. Movable 15 to 20 degrees. Spine ossified: vertebræ immovable. Scoliosis toward left 150-160 degrees, in cervical region lordosis almost 90 degrees. Musculature of back ossified and united to scapulæ. Gluteal insertion ossified. Left trochanter major has exostoses 7 cm. in diameter. Adductores femoris ossified and aponeurosis hard. Tensores fasciæ hard. Vastus intern. on condylus intern. ossified. Right knee almost ankylosed at 90 degrees. Exostoses in right popliteal region and in pes anserinus 4 cm. long. Triceps suræ hard as though ossified. No microdactylia mentioned.

Case C.—Reported by Author R. Elliot, in 1911. Female, aged 17 years. Family history neg. First symptoms appeared shortly after second year—lumps appeared on her head and disappeared again; no apparent cause, no injury. In the succeeding three years more nodules formed. At 5 years torticollis appeared and remained for about six months. Later painful swellings on arms and legs interfered greatly with motion. Since that time patient has never been able to abduct her arms or raise them. A valvular heart lesion was then discovered; no previous acute infection or rheumatism. January, 1907, typhoid; July, 1907, stiffness of jaw; teeth could be separated but slightly; hard bony mass fills submaxillary region. Hard circumscribed swellings free from ribs on both sides of thorax. October, 1907, both arms became stiff and partial fibrous ankylosis developed in right elbow-joint. Left shoulder and arm followed. Head bent forward and downward; stiff. Maxillary movements much restricted. Teeth separable only about one inch. Lower jaw displaced

to right side. Anterior and posterior neck muscles firmly indurated. Mastoid part of sternocleido of bony hardness; clavicular origin indurated. Pectoral muscles hard and contracted. Hard nodule on upper border of left scapula. Small exostoses on right eighth and eleventh ribs; also ninth left; also on posterior crest of iliac bone. Body bent forward; rotation of trunk impossible. Spine firmly rigid. On either side of lower dorsal and lumbar spine a broad bony mass, over right inguinal region a firm induration size of a man's palm. Abdominal muscles tender to pressure. Muscles of legs free, but patient states that they feel too short. Both thumbs show bony ankylosis in terminal phalangeal joints. Second joint pliable, but atrophic. Thenar flat and atrophied. Never had voluntary movements of thumbs. They are held flexed on palms (pollex valgus). Microdactylia of both big toes; absence of one phalanx (?). Toes directed outward, lying partly under second toe (hallux valgus). No. X-ray of toes. Cause of short-hallux probably erroneous, and shortened metatarsus primus; here the usual explanation—X-ray of hands shows shortened metacarpus primus.

Case CI.—Reported by Jgn. Peteri and Gust. Singer. Male, aged 4 years. Family history neg. First symptoms appeared at 1½ years of age—swellings on right and left sides of forehead. They disappeared again after a few days. Other swellings appeared, which grew harder and finally became as hard as bone. 1906: head bent forward and fixed rigidly, only slight lateral motion. A swelling size of a child's fist developed in submental region; during his hospital stay other inflammatory symptoms. With relief of these symptoms a solid mass remained, impeding movement of lower jaw. Ridges of bony substance in temporal and masseter muscles. Muscular part of nape forms a diffuse, solid bony mass with nodulated surface, causing rigid stiffness of neck. Slender bony ridges in both sternocleidi bound to broader ones in deeper cervical muscles. Pectorales major and minor changed to hard osseous plates. The upper arms are firmly united to shoulder-blades and ribs by broad hard bridges, in anterior and posterior axillary folds. Both biceps and brachialis ant. contain broad bony ridges along entire length. While the right one is partially fixed to humerus, the left runs free from bone to tuberosit. ulnæ. Both scap. fixed to back. Musc. of forearm tough and fibrous. Entire vert. column rigid. The processi spinosi form, together with muscles and ligaments, thick, heavy, protuberating masses of bone tissue Four years later: patient again admitted. Stiffness and immobility of entire upper part of body. Angular conformation of shoulder girdle and various relief-like elevations throughout body had increased, especially those of obliquus intern. Microdactylia of both thumbs and big toes. (Synostosis between short first metatarsal bone and first phalanx.)

Case CII.—Reported by Felix Bauer, in 1911. Aged 2½ years. Sex not mentioned or alluded to in entire paper. Family history neg. Scarlet fever when 6 months old (?). First symptom noticed was ossification of neck after scarlet fever. Ossification of neck and shoulder progressed and caused decrease of movability of head and arms. Strained, bent-forward posture of head and upper part of body. Upper extremities fixed. Spinal column stiff. Sternocleidi and the deep neck muscles, the shoulder muscles and both bicipites are ossified and intersected with projecting lumps and spurs. They are mostly fixed to the underlying bones; only those of the biceps are free and movable in the muscles. From costal arch in anterior axillary line extends a round partly fibrous and partly osseous ridge size of penholder toward the symphysis. It continues upward toward the fifth rib on both sides. Over the shoulder-blades and at sides of spine several immovable, semiglobular, osseous tumors. Microdactylia of both big toes; in a lesser degree of both thumbs and little fingers.

Case CIII.—Reported by Otto Jüngling. Male, aged 6 years. Family history neg. First symptoms appeared before end of first year—lumps developed in nape

of neck. At first lumps disappeared again without leaving any traces, but after some time they recurred in the same place, remained and grew hard. Gradually this process spread from the neck to the back and arms, and for some time the upper part of the boy's body has been stiff. Head, neck, shoulders and trunk quite stiff. Mouth can only be opened 1¾ cm. In the neck only the vert. prominence can be felt; from it a bony ridge 4 cm. long, thickness of a small finger, starts downward, divides into two and at level of scapula turns up again. At about the ninth vertebræ another ridge starts obliquely to the angle of the left scapula. On the right side only two lumps. The lumbar portion of the spine shows slight scoliosis convexity to right. The sacrospinalis on the left is stone-hard in its entire length. A small bone plate in the height of the fifth lumbar vertebra. The upper arms are fixed to the thorax in an angle of 35 degrees. From the scapula angles hard ridges run to both humeri. In both triceps are hard bony masses and two stalactites in the right elbow head. Both thumbs very small, due to smallness of first metacarpus. The first and second phalanges of second and third fingers very small and ankylotic. Shortness of right first, third and fourth and left first, fourth and fifth metatarsal bones. The left first, fourth and fifth metatarsal bones show only minute epiphysial nuclei (distal) and the phalanges of all the toes are deficient. The first metacarpal metatarsal bones show only proximal, the other second, third and fifth only distal epiphyses. Hypospadia-urethral orifice at scrotal insertion.

Case (?).—Reported by Riehl, in 1912. Male, age (?). No family history. First symptoms appeared since 6 years of age—febrile attacks of pain in joints, with redness and swelling of skin. Later lime deposits in muscles and subcutaneous tissue. Skin difficult to fold; thickened in places, in others atrophic. Very little fat in subcutaneous tissue. Irregular and indistinct cloudy or striped shadows and very little pronounced ossification in X-ray picture. Author speaks himself of similarity of his description of this case with severe sclerodermia. (Not myositis ossif. progress.) No mention of microdactylia.

Case CIV.—Reported by C. A. Manjapa, in 1912. Male, aged 25 years. Family history neg. Feverish illness at 6 or 7 years of age. First symptoms appeared at 6 or 7 years of age—had fever for nearly six months, and about that time lumps were discovered on back of neck and chest. Grew gradually unable to stand erect or walk freely and became mentally deficient. His disease, on account of lumps, was erroneously diagnosed as leprosy and he was transferred to the leper asylum; later, in September, 1907, on account of his mental condition, to the lunatic asylum. Patient can only sit and stand in a crooked position. Head fixed, with deflection to left side; incomplete lockjaw, due to stiffness of masticating muscles. Neck stiff and almost all voluntary muscles in slight or greater degree of ossification. Trapezius and erectores spinæ present, bony ridges extending from suboccipital to lumbar region and bony tumors at spine and inferior angle of right scapula. Left pectoralis major hard; bony ridges along sternal fibres, connecting with one from their humeral insertion. Right deltoid also shows bony ridges. Right forearm fixed at right angles to arm by bony buttresses. Left forearm fairly movable, but not to full extension. Right supinator longus entirely ossified and bony swellings in flexors and extensors of left forearm. Left thigh has movable bony plates in region of extensor quadriceps, adductor longus and hamstring muscles. Had short fingers and short toes. About eight months prior to his death (July, 1910), from dysentery, patient began to lose flesh and strength. The joints became more firmly fixed and the osseous growths very prominent and distinct.

Case CV.—Reported by Fritz Magyar, in 1912. Female, aged 3 years and 9 months. Family history neg. First symptoms appeared at 3 years—hindrance in motion of muscles of upper arm and pains on pressure. Pains disappeared, but hindrance remained and muscles grew stone-hard. Head bent forward. Ossification of both

bicipites brachii; on back also ossifications in serrated form adhering to underlying tissues. The movements of arms were interfered with by hardening of upper and lower margins of latiss. dorsi posteriorly and pectorales major anteriorly. No mention made of microdactylia. (Brief notes only in Wien. mediz. Wochenschr.) To personal letter received no reply.

Case CVI.—Reported by David Rankin, in 1912. Female; a little negro girl. Subsequent course of disease unknown. Casual observation of author during visit to a hospital in Brazil, where he saw a little black girl suffering from myositis ossificans progressiva, with trapezius, latiss. dorsi and greater portion of both pectorales ossified. No mention made of microdactylia.

Case CVII.—Reported by Joseph Frattin. Female, aged 14 years. Family history neg. First symptoms appeared at 1½ years of age—painful swelling at nape of neck; diasppeared again within twenty days without leaving a trace. At 4 years painful swelling again developed in same place and disappeared again, but neck remained stiff; later back and hip, which likewise showed exceedingly hard tumors. She suffered from epilepsy at an early age. The attacks grew rarer, and two months after leaving hospital disappeared entirely. Head slightly inclined toward right; can be turned, but not freely moved from side to side. Hard nodules on both sides of glabella, some adherent to bone. Muscles on both sides of nape of neck have bony-hard nuclei at occipital insertion, the right sternocleido at mastoid and a bony thickening at clavicular. Lower insertion of scaleni ossified. Upper margin of cucullaris and upper part of left sternocleido ossified. Tendon changed to osseous band. Both arms fixed in adduction. In right biceps and triceps, pectorales, latiss. dorsi, many bony plates; on left side tendons of these muscles ossified; on lower epiphyses of humerus irregular bony mass encircling joint completely. Breathing abdominal. Broad muscles of back completely ossified, left more so than right. Infraspinatus and sacrolumbalis show osseous plates movable in muscular tissue. Near sacro-iliac symphysis are osseous tumors; both hips stiffened; soft parts surrounding coxo-femoral joints show many ossifications. At lower part of glutæus major large osseous band and an osteoma on inner side of crista ilii. Similar conditions on left leg. Head, slightly inclined toward right, can be turned but not freely moved from side to side. Both big toes show microdactylia. Left smaller than right. X-ray picture not quite clear.

Case CVIII.—Reported by S. Goto, in 1912. Male. Family history neg. First symptoms appeared at 1 year and 10 months of age—swelling and hardening in jugular fossa. Later an osseous formation developed in the jugular swelling. A year afterward swellings on occiput and various other parts of the head, which disappeared again. Since January, 1911, parents have noticed stiff posture of body. Weight, 14.525 kils.; height, 93.0 cm. Movements of head very limited. From posterior surface of mandibula comes a hard osseous band to posterior surface of chin. Small tumors at sternal origin of sternocleido and at insertion of trapezius. Both scapulæ fixed to thorax and their lower angles are united in one solid mass with surrounding ossified tissues. On both sides of sixth to eighth dorsal vertebræ bony growths size of thumb. Along both sides of spine broad, symmetrical, flat, bony growths 3 cm. broad, united below to crista ilii, above to masses at sides of vertebræ. The ant. and post. axillary folds are ossified. Arms only very slightly movable in shoulder-joints, free in elbow and other joints. A string-like band in right side of abdomen and a small, hard, osseous thickening on inner condyles of femur and tibia. Both thumbs and big toes abnormally small, owing to smallness of metacarpal and metatarsal bones. (Hallux valgus.) Electrical sensibility; decrease to absence of irritability to faradic or galvanic current in affected muscles. Those apparently free from affection show no disturbance.

Case CIX.—Reported by Rudolph Jacobi. Male, aged 6 years. Family history neg. First symptoms appeared at 3 years of age—hard nodules formed on back.

Nodules were rather flat at first, but grew gradually to their present size. For the last five weeks there has been a disturbance of the free motion of his right arm. Movements of head not free; chin cannot be brought to thorax. Scattered over entire back are numerous osseous protuberances of semiglobulous and flat form, averaging pigeon-egg size; also long ridge or beam-like elevations of about 15 cm. length and thickness of thumb diverging from median line to lateral parts of thorax. Nearly all the muscles of the back are changed into a hard osseous mass, encasing the ribs armor-like and inseparably connected with them. Respiration abdominal; thorax shows only minimal excursion. Arms and legs show no change except exostoses on both condyli interni tibiæ, 2 cm. long. Microdactylia of thumbs and big toes. (Hallux valgus.)

Case CX.—Reported by S. J. Khaikis, in 1913. Male, aged 4 years. Family history neg. First symptoms appeared when about 3 years of age—old swellings appeared on occiput. Later swellings showed on back and lower extremities (treated by local physicians with inunctions); they would subside, then reappear. These tumors began to appear first on the occipital musculature, then on that of the back and upper ribs. The swellings along the ribs were quickly absorbed, but the others hardened and had the consistency of bone. Patient only complained when new swellings appeared. Boy normally developed. Pain when bent over, but head can be bent forcibly in the different directions without pain. Occipital movements interfered with by tubercles on occipital bone about attachment of cucullaris. Similar tubercles on both sides of spine along lower third dorsal and first lumbar vertebræ. Other nodules appear over middle of upper ribs; also in axillæ, interfering with raising of arms. During observation of one month methodical treatment with potassium iodide, baths and fibrolysin. Condition grew worse, redness and swelling to left side of second lumbar vertebræ. Redness disappeared again, but swelling grew osseous. No mention of microdactylia.

Case CXI.—Reported by Volhynia Medical Soc., 1913. Female (?). Subsequent course of disease unknown. The foregoing case was before his discharge shown in the Volhymia Medical Society. A similar case of a girl was demonstrated there also. Demonstrator not mentioned in report.

Case CXII.—Reported by Erich Blenkle, in 1914. Male, aged 21 years. Family history neg. Not much known about early history; lost parents very early in life. Subsequent course of disease unknown. Stature of boy 10 to 12 years; height, 137 cm.; panniculus underdeveloped. All the limbs except the right leg and hand stiff; cannot sit down, has to do his work, eat, etc., standing. Impossible to mount stairs. Jaws cannot be opened more than ½ cm. Teeth (incisors) have been removed for feeding. Masseters hard, with hard osseous nodules imbedded. Head only movable sideways in very slight degree. Spine completely stiffened. Sterno-cleido and neighboring muscles of neck, entire musculature of back stiff and hard and interspersed with small osseous nodules. Along entire dorsal spine large osseous plates; below tenth and eleventh vertebræ no muscles to be felt; only one continuous osseous plate, extending to crista ossis ilii. Thorax and ribs immovable; breathing entirely abdominal. Pectorales and deltoids stiff and hard. Both arms fixed, left less so. Osseous imbeddings in biceps and triceps and near origin of extensor carpi radialis. Slight volar flexion possible in left wrist. Both hip-joints immovable; hard masses surround both trochanters; knee-joints slightly movable; osseous plates in left adductor. Exostoses of femur; hard mass around left knee muscles of right leg, around hip and knee-joint hard and stiff. Both thumbs small (micro-dactylia). Third phalanges of fourth and fifth fingers turned toward radial side. No microdactylia in either foot.

Case CXIII.—Reported by F. Parkes Weber and Alwyn Compton, in 1915. Female, aged 2½ years. Family history neg. First symptoms appeared at 7½ months of age (March, 1913)—was shown before the Royal Society of Medicine, with bony

projection from left side of neck and thin spicula of bone attached to middle of the back of left clavicle. In March, 1914, various diffuse swellings appeared, involving muscles and superficial fasciæ; skin could be freely moved over them. They disappeared and soon were followed by others in left scapular region and left lower part of thorax. Similar ones appeared also on right side. In May, 1914, a distinct bony formation appeared in left posterior axillary fold, and transient swelling in right pect. maj. In June hard swellings in right biceps and lower part right scapula and corresponding ones on left side. In September and October a bony lump on occipital insertion of trapezius. Very stiff in movements of back and neck, extension grows more and more limited. There also seems to be osseous infiltration in both anterior and posterior axillary folds, limiting abduction of both arms. Microdactylia of both thumbs and big toes; in the thumbs due to smallness of the first metacarpal bones; in the toes to a synostosis and resultant smallness of phalangeal bones. (Hallux valgus.) A small piece of right latiss. dorsi examined microscopically showed invasion of muscle by newly formed fibrocellular connective tissue.

Case CXIV.—Reported by Angel M. Centeno. Male, aged 8½ years. Family history neg. At 3½ years phlegmonous (?) inflammation of left sternocleido. First symptoms appeared at 6 years of age—entered Children's Hospital. Smooth, hard, painless tumors in region of right scapula. (Caught measles in hospital, but recovered without complications.) When seven months in hospital first operation was performed on sternocleido tumors. Result not known to padres. Later new, round, hard, painless tumors of various sizes appeared over the entire region of the dorsalis major, the trapezius and the costal walls. Shortly after the first operation a second one was performed, and a little later a third one, for the avowed purpose of removing little pieces of new-formed bones. Wassermann positive. (Author seems to attach much etiological importance to this reaction; in other such cases reaction was negative.) Face looks as if patient has adenoids and prognathism of left maxillary bone. Head inclined forward and downward. The physiological lordosis has disappeared. Thorax slightly inclined forward; arms fixed in the same direction as upper part of thorax, separated from it and in attitude to grasp an object. Osseous tumors in dorsalis longus, trapezius and both sacrolumbales; in fact, entire posterior wall of trunk. In upper movement of left trapezius a perfectly movable, painless, small, hard nodule. Lower extremities slightly flexed, left forming an obtuse angle with thighs. Double hallux valgus turned outward and metatarsophalangeal at nearly right angles with first metatarsal bone, covering with the lateral edge of the phalanges the second metatarsophalangeal articulation. There is a disturbance in the development of the two upper and inner incisors. They show an excrescence, which descends to the point of an implantation from the inner side to the middle of the teeth. The two rows of teeth do not meet together, and in the hard palate is a deep oval impression. The voluntary motions are limited by the described lesions. Left arm passes with difficulty just the horizontal line; right one appears as if glued to the side, its angle of motion not more than 30 degrees. Hands show thinness of little fingers and shortness of thumbs at expense of last phalanx (?). No X-ray of either hands or feet. Both testicles are ectopic, having remained in the inguinal canal near the external opening. Nothing else of note.

Case CXV.—Reported by Seth Hirsch and Joseph Roth, in 1917. Male. Family history neg. First symptoms appeared at 2 years of age—lump on right side of neck. Lumps on neck disappeared. Other lumps developed below right scapula. Said in orthopedic hospital he had suffered from tuberculosis, applied a brace for nine months. It was then removed. Incision was made upon lump below right scapula. During last year the shoulders were becoming progressively displaced forward. Tonsils and adenoids were removed a year ago. Slimly built; walks about, but is constrained in movements of back and arms; shoulders very narrow, as though cramped forward. Calvarium shows at right parietal region large soft tumor about

two inches in diameter, seemingly set into the bone. Both scapulæ are tilted forward and seem to be fixed in relation to the ribs. A bony protuberance below right scapula. Some pain at movements of shoulder beyond difficult limitations. Very rigid spine throughout. (Pokerback.) Normal genitals. Shortenings of both thumbs and big toes. X-ray shows below the angle of the left scapula a large, rounded bony mass, and extending upward and inward a strip of bone located in the teres major. At the right side a large bony mass posteriorly behind the tenth and eleventh ribs, extending upward and outward into a rib-shaped prolongation, bounding the inferior axillary space and divides near the humerus into two small branches. This entire ossification is in the latissimus dorsi. In the right lumbar region opposite third and fourth lumbar vertebræ is an irregular stellate ossific shadow, and extending upward a linear mass merging with the body of the twelfth dorsal. On the left side runs an ossification strand from the lower ribs toward the iliac crest. Knees show a spur-like, bony formation from the posterior surface of the lower and of the left femur diaphysis. Also small ossification strand above its head. Thumbs and big toes show brachydactylia.

Case CXVI.—Reported by Seth Hirsch and Joseph Roth, in April, 1917. Family history neg. First symptoms appeared at 6 months of age—small nodule on forehead, increasing in size to that of a plum, then remained stationary. Well after lump on forehead till age of 8 years, when it was noticed that he could only raise the right arm with great difficulty. Soon after the left became similarly affected. Patient was removed to post-graduate hospital, where incisions were made over both scapulæ and pieces of a bony growth removed. Child got scarlet fever; returned home. Conditions progressed slowly, involving neck and rest of the spine. Later the lower extremities became also involved; boy still being able to walk with some difficulty. The disease shows signs of progression. Patient very thin, mentally active, able to walk with difficulty. Spine rigid and kyphotic, neck rigid, arms are adducted; he is unable to move them either outward or upward. Scars over both scapulæ from which portions of the osseous growth have been removed. There is a bony growth about the left frontal bone. The posterior muscles of the neck appear hard to the touch. The scapulæ are markedly winged. The upper extremities are fixed to the axillary border of the thorax. There are hard bony masses running from the serratus magnus on each side to the humerus. Posterior spinal muscles appear hard; there is marked rigidity of entire spine. There is marked contraction of the adductors and the tensores fasciæ latæ. The thighs are flexed on the pelvis, fixed and adducted; the legs are flexed on the thighs. Hard bony masses in the adductors and muscles of the calves. The genitals are normal. The big toes are considerably shortened. X-ray examination. Numerous strands of ossification in both latissimi dorsi and in many of the intercostal muscles of both sides. Examination of the pelvis shows numerous branch-like strands of ossification extending into the left gluteal group. A large spur-like mass of bone between the lesser trochanter and the right ischium. The knee shows a long spicule of bone springing from the posterior surface of the femur. Both big toes show brachydactylphalangia.

Case CXVII.—Reported by Eug. L. Opie, in May, 1917. Male, aged 70 years. Nothing mentioned of previous history, description of skeleton only given. Subsequent course of disease unknown. Left internal pterygoid muscle ossified. The anterior edges and articular surfaces of the cervical and lumbar vertebræ show bony projections. Head and neck of seventh and eighth ribs ankylosed by bony union with the corresponding vertebræ. Broad bands about 2 cm. across connect on the right side of the fifth, sixth and seventh ribs about 2.5 cm. from their vertebral origin; the fourth and fifth ribs are similarly united on the left side. The bones of the shoulders and upper extremities show prominent ridges for the insertion of the muscles. No deformity of hand. Pelvis shows similar prominences and projections also on edges of acetabulum.. Both femora show irregular ridge of bone 1 cm. high on the linea aspera just below the middle of shaft.

Tibiæ and fibulæ very irregular in outlines. Rough areas with spiculæ project upward in the direction of the muscles. Bones are considerably thickened by periostitis; membranæ interossei are ossified at irregular places, forming ridges of bone. Right foot: porous texture of bones. Distal end of first metatarsal is enlarged and irregular in contour. Projecting downward and backward from the head of bone is irregular osteophyte 1 cm. long. Small bony projection, 3 cm. long and ankylosed, at the upper inner surface of the articular surface representing the two terminal phalanges of the great toe. These bones do not extend beyond the first interphalangeal point of second toe. Left foot: shows some porous texture of bones. Head of first metatarsal bone absent; end of bone broadened. Appearance of metatarsal bone as if its head, together with big toe, has been amputated. Microdactylia of both big toes (?).

Case CXVIII.—Reported by Ten. Bokkel Hunink, in June, 1917. Female, aged 7 years. Family history neg. First symptoms appeared at 2½ years of age—swelling under chin. Swelling under chin disappeared; was followed by one in neck, causing stiffness. Had hard nodules on forehead, which also disappeared later. Mouth can only be opened ¾ cm. Flexion in elbow-joint to 90 degrees, extension to 120 degrees. Left shoulder hardly movable. Along the upper ribs many bone-hard swellings. The lower third of the left m. biceps feels like a bone-hard mass. The left axilla is entirely enclosed with bone-hard plates; no muscle to be felt. The muscles of the neck, with the exception of the sternocleidomastoidei, are changed to a bone-hard mass. Large bone-hard plates in the long muscles of the back; also in the left latissimus dorsi. The medial edge of the right scapula merges into a bone-hard mass, which is fixed to the boneplates in the long extensor muscles of the back. Returned later with about the same status, only that the mouth can be opened wider and the muscles of the neck, which felt like a bone-hard mass at the first examination, now have grown softer, and though not as soft as ordinary muscles, no bone-hardness can be felt in them, and the movability of the head is accordingly improved. X-ray plates not shown in this publication, but author mentions that only in the chest and back the palpatory findings are confirmed by the X-ray, while the neck muscles show no ossification and the biceps show only a very small piece of bone in the muscular tissue. Bilateral microdactylia of both big and little toes; big toes have large first metatarsus and only one very small phalanx. (Synostoses?) Little toes very small phalanges.

Case CXIX.—Reported by C. Johannesen, in 1917. Female, aged 2 years and 8 months. Family history neg. First symptoms appeared at 1 year and 8 months of age—swelling over skull and occiput. (Protub. occip.) Swellings were not sensitive. After four months later swelling over left shoulder, spreading to left scapular region and left side of neck to ear. It traveled in four to five days under left arm and across back. Not sensitive; no discoloration of skin. Arms could hardly be moved in shoulder-joints. After a week swellings began to disappear; instead a swelling appeared in left sternocleidomastoideus. After the left-sided swellings had disappeared swellings showed in right scapular region under right arm and lumbar region. About two weeks ago patient became sick; had chills; took to her bed and has lost flesh. Patient well developed. Height 88.5 cm., weight 13.8 kg. Head circumference 52, chest 54 cm.; anæmic; purulent discharge from both ears; nasal discharge and cough; chest and abdomen normal; temp. 37.2; pulse 92, regular; sits rigid; bends head forward and over to right side, with face toward left; opens mouth wide and masticates and swallows without difficulty; right side of neck and right fossa supraclavicularis are swollen; swelling of lower half of sternocleido. The inspection of right side of back shows swellings and grooves like topographic chart. Swellings in the neck in fossa supraclavic. continue over right part of back down to eleventh rib. Swellings in muscles of trapezius, axillary folds and serratus ant. They reach middle axillary line from second rib down to eighth rib. Swellings are bone-hard and immovable over underlying tissue. Skin normal appearance. Right arm

stiff; left can be moved freely. Wassermann and v. Pirquet neg. Erythrocytes, 6,000,000. Leucocytes, 125,000. Urine nothing abnormal. Microdactylia of both thumbs and big toes. Hallux valgus bilateralis. According to Roentgen plates the thumbs show a shortened first metacarpus and both big toes a synostosis of a shortened first metatarsus, and first phalanx with a separate small last phalanx outward, resulting in bilateral hallux valgus.

Case CXX.—Author's own case, in 1916. Male, aged 20 years. First symptoms congenital. Subsequent course of disease reported elsewhere in this text. Status at time of observation reported elsewhere in this text. Congenital abnormalities reported elsewhere in this text.

Author's Case

L. B., newsboy, born 1896 in Rumania; normal confinement. Family history negative; both parents alive and well but very ignorant, and the history (previous to his coming under our observation), having been extracted from them under great difficulties, is not as complete as I should wish.

Two brothers (nine and twelve) and one sister (fifteen years) in good health. No T. B. or history of any other systemic disease in immediate family. Immediately after birth the midwife noticed a tumor in the right groin the size of a small egg. This tumor probably contained bony spicula; as the mother, who gave the details of the previous history, describes it as having felt to the touch as though filled with nails. His right leg was also shorter than the left. The attending physician advised a watchful waiting and to postpone surgical interference for one year.

At the end of that time the tumor had grown down the thigh to the leg, while the portion in the groin had grown to the size of a fist. He was operated on at the age of fourteen or fifteen months by a Rumanian surgeon who removed the tumor in the groin, leaving the remainder until the child had grown older and stronger. He stayed several months at the hospital. The parents, somewhat later, emigrated to the United States and at the age of five years patient was placed in the New York Orthopædic Hospital for a second operation.

An incision was made from the lower third of the thigh to middle of leg and a part of the tumor apparently successfully removed. The functional result was not improved, the leg remaining stiff, neither flexion nor extension being possible.

After three years patient again entered a hospital and remained there another eight months. It is not clear what was done at that time; parents say that a tumor in the upper part of the leg was excised. No further operation was done until he presented himself in my surgical Out-patient Department of the Mount Zion Hospital, under the care of my then first assistant, Dr. A. Newman, now Professor of Proctology C. P. & S., in the latter part of 1908. He complained of pain in the left upper arm and over his right tibia below the knee.

His status at that time was: A rather slender boy, good color.

Head and neck nothing abnormal. Left arm shows bony ridge in and under deltoid muscle, extending from below deltoid process and

along anterior outer surface of humerus to insertion of deltoid tendon. Humerus thickened; patient cannot raise his arm on account of pain when attempt is made to bring deltoid into action. Both thumbs show very short and broad last phalanx.

In the right groin a scar and another extending over inner part of right thigh, both from former operations. Exostotic growths spring from crest of right iliac bone and along the line of Poupart's ligament. There are small isolated isles of bony masses on the abdomen in the right iliac region, extending into right obliquus and rectus abdominis half way up to the umbilicus.

The entire adductores region of the right thigh, from their origin down to the lower third of the thigh, stonehard. The anterior part of thigh between lesser and greater trochanter also forms a bony mass; the femur is thickened; as the mass nears the knee it swings backward and entirely fills the popliteal space. Knee completely ankylotic. The mass then passes down along the inner side of the leg into the foot, running along the inner margin of the latter to the big toe. Microdactylia of both fourth toes.

Priessnitz compresses were ordered, to be changed every four to six hours. The pain disappeared soon after they had been regularly applied.

Entered hospital on August 31, 1909, with pain in left humerus and lower epiphysis of left ulna, and over right tibia below knee. Attack accompanied by fever. X-ray of left arm and hand shows thickening of upper two-thirds of humerus and periosteal thickening along lower part of ulna. In humerus just above epiphysis, oval wedge-shaped space giving impression of cavity. Tuberculin test negative. Under treatment with warm Burow compresses pain gradually disappeared but left thickening along ulna. Discharged October 2, 1909.

He was not seen again until the latter part of September, 1910, when he came into the clinic complaining of severe pains in the left upper arm. It was swollen, and painful to pressure and percussion all along the upper two-thirds of the humerus, and the skin appeared slightly œdematous. The pain had begun about a month before, but his great fear of a possible operation had kept him away from us until the pain had grown too violent to bear. Rest, ice and diluted Burow solution compresses were first tried, upon the urgent request of the boy, but gave no relief, and on October 2, I made an incision over the anterior and outer aspect of the humerus through the fibres of the deltoid, from the shoulder downward along the upper two-thirds of the bone. In the region of the insertion of the deltoid tendon the bone was bared, and on finding considerable roughness, an opening was made into the medullary cavity. A small quantity of pus was found in the medullary cavity (two cultures taken), together with considerable granulation tissue. The upper two-thirds of the cavity of the shaft were opened and curetted clean, until normal tissue below and above was found. The cavity was dried with the Hollander hot-air instrument and an equal part mixture of carbolic acid and tincture of iodine applied over entire exposed surface, after all bleeding

had been stopped. A paste of plaster of Paris, composed of sterilized plaster in 1 to 500 cyanide of mercury solution to fill the entire cavity is put in and allowed to harden. Periosteum and muscle are united with interrupted catgut, and skin with continuous silk sutures. Uneventful recovery. (Culture staphylococcus; no T. B.)

In the latter part of April, 1911, the patient came to us again complaining of considerable pain in the right iliac region during locomotion but not while at rest. Examination shows lower 5 cm. of right rectus muscle ossified. Mass very hard, a sharp-edged piece of bone raising skin tight.

May 13th: Incision 5 cm. long parallel to Poupart's ligament and about 1½ cm. internal from it near insertion of right rectus muscle. Muscle here completely ossified and a spur 2½ cm. long and 1½ cm. thick protruded down into the tendinous part, but entirely free from os pubis. It was removed, edges of muscle smoothed and wound closed. Discharged May 23, 1911.

A small piece from the lower edge put in trichloracetic acid for decalcification and microscopical examination.

On August 15th, 1911, returned to hospital. A large spur had formed, extending from the right anterior iliac spine to the edge of the obliquus externus of that side about 5 cm. long and 3 cm. wide, causing considerable pain. Incision over spur; blunt separation of adherent muscle and chiselling off of spur from anterior iliac spine. The rough bone edges smoothed off and hæmostasis with hot saline solution. Fascia united with chromic catgut; skin subcuticular. Uneventful healing; discharged August 30, 1911.

Returned in September, 1912, complaining about two closely neighboring ventral herniæ in the right iliac region where previously osseous plates had formed and which had been extirpated. The muscular fibres had atrophied and the ventral hernia developed. An attempt was made to bisect longitudinally a piece of the sound upper part of the right rectus abdominis muscle into an anterior and posterior half, and having tied off and resected the peritoneal sacs, turn the anterior flap down over the hernial apertures and fasten it to the lower tendinous part. The result was not very successful; part of the muscle-flap sloughed and the defect in the abdominal wall was only slightly improved.

About three weeks after operation the patient's face and both arms became œdematous and a marked hæmaturia appeared. The face and arms resumed their natural condition after two days, but the hæmaturia remained for several weeks. It finally cleared up without leaving any trace, nor has it ever returned since. (Late ether nephritis?)

A cystoscopic examination was stubbornly refused by the patient. Urine contained at first some granulated casts together with blood. After a few days casts disappeared. No pain or dysuria.

We did not see him again until May 26, 1914. As the status since then has remained about the same, I will give the one taken at that date.

Status.—Good complexion; red cheeks; looks like a boy of 15 years. Is underfed; weighs 113 pounds; rather infantile habitus; voice harsh and deep.

Head well-shaped and covered with an abundance of black hair. Face smooth, no sign of beard. Eyes clear, pupils equal, dilated, react to light and accommodation. Mouth: Mucosa normal, tongue clear; teeth carious, gums show signs of pyorrhœa alveolaris.

Pharynx inflamed, tonsils normal. Neck long, slender; no general glandular enlargement; thyroid gland not palpable; no visible pulsating blood-vessels.

Chest: Sternum in depressed groove, insertion of ribs on both sides forming convex arches to body and ensiform cartilage (ratite or ecarinate chest); entirely free from hair, which is also absent in the axillæ. Excursion on normal respiration is not marked. Breathing is mostly abdominal. On deep breathing, however, thoracic movements are extensive. Expansion is about 5 cm. Mobility on both sides equal.

Palpation: No muscular resistance; T. F. somewhat exaggerated on right side. Percussion and auscultation normal, slight broncho-vesicular breathing below second and third interspaces. Slight lateral curvature.

The left arm shows a scar from the acromial process to the middle of the humerus, on its anterior and outer surface. The bone is thickened and rough and of irregular shape up to its middle, where it again assumes its normal size and contour. The change in form of the upper part consists of a nodulated groove 3 mm. deep on its anterior and outer surface. In the right iliac fossa, beginning about 3 cm. below level of anterior superior iliac spine, free osseous plates can be felt in the rectus and external oblique muscles. The skin is freely movable over them, but they are firmly imbedded in the muscle. Hardly any hair in pubic region.

From the right horizontal branch of the pubic bone an exostosis starts upward in the substance of the obliquus muscle for about 5 cm.

Its base is firmly attached to the os pubis; it is about one-half a ctm. wide and grows gradually wider toward the tip, where it is 2½ ctm. Near the tip a free osseous plate, at the edge of the right rectus, is movable and moves with respiration. This plate is about the size of a large melon seed. There are two smaller exostoses, one on each side of the large one, firmly connected with the pubic bone; the outer one, extending down to the large ossification of the thigh, connected with the femur; the inner one, near the symphysis, nearly joining the loose plate in the rectus above it. This exostosis is about 3 cm. by 1½ cm.

Between the outer and middle, also between the middle and inner exostosis, the muscle and skin are atrophied in two places and a ventral hernia protrudes through each of them, giving a marked visible and palpable impetus upon coughing.

The right thigh is ankylotic in hip joint and is flexed at an angle of about 35° and adducted. The knee is also ankylosed and in flexion at angle of about 25°.

The right leg shows ossification of the entire adductor and hamstring muscular groups; extensors and abductors not so well developed as on opposite side.

Circumference of middle of thigh—right, 30 cm.; left, 47.5 cm. Circumference just above patella—right, 28 cm.; left, 32.5 cm.

Nearly the entire mass of the inner and posterior muscles of the right leg is ossified; except the extensors, the peronei, the tibialis anticus, and the lower part of the soleus.

Circumference of calf—right, 24.5 cm.; left, 31.5 cm. Length of leg—right, 67 cm.; left, 80 cm.

Flexion of right foot is very limited, but movements of tendon Achilles are felt very distinctly. The foot is in adduction and its inner two-thirds ossified up to middle of metatarsal bones. The fourth toe of each foot is small and bent across third toe.

Patient complains of pain in the right lumbar and inguinal regions, particularly severe when standing erect. This pain commenced in a slight degree about eight months ago and gradually increased in intensity and duration, becoming lately so severe that patient was compelled to give up his work and apply to the hospital.

Examination: Shows small spur of bone in the right inguinal region about the middle of Poupart's ligament, about 1½ centimetres long and pointing towards horizontal branch of the pubic bone. Another sharp spur is on inner side of the femur, to the inside of crural canal.

Operation (May 28, 1914).—Incision over both spurs; the one in the abdomen can be easily removed by excision. Attenuated muscle and fascia are sewn together with catgut and skin with Michel clamps. The spur on the inside of the femur lies directly under the skin, and after incision it is removed with chisel; after hemorrhage has been stopped the skin is united with Michel clamps. Both wounds healed by first intention. Patient dismissed June 8th.

Returned to hospital October 25, 1915. Patient has been perfectly well ever since leaving hospital. About ten days ago he began to have pain over posterior surface of arm just above the wrist. This is a sharp sticking pain which comes on on movement of wrist or contraction of muscle of forearm. Greatest pain is when he flexes the wrist. Appetite good, bowels regular.

Examination reveals small highly sensitive exostosis, about the size of a small melon seed, over lower end of radius on dorsal surface; small hard nodule in *skin* above extensor tendon two inches above the wrist. Nodule is freely movable with skin and sensitive. Similar nodule over middle of left calf; underlying muscles and entire posterior part of left leg free from any other ossifications.

Heart and lungs normal.

A coagulation test of patient's blood was made, found normal; time 5 minutes.

Operation October 27.—For removal of exostosis on posterior surface of radius two inches above wrist joint. Esmarch's bandage applied to hand and arm, up beyond elbow. Local anæsthesia with novo-

caine. A longitudinal incision was made through skin, and subcutaneous fascia tendons of extensor muscles separated, deep fascia and periosteum incised and the latter levered from bone with elevator and pulled aside. The bared exostosis was then removed with bone forceps and base smoothed off with chisel. The small nodule with a small oval of surrounding skin was then removed under local anesthesia for microscopical examination, also that on left calf. Wound over wrist closed in two layers, deeper layer with interrupted catgut sutures; all skin wounds closed with Michel clamps. Dismissed October 31, 1915, after uneventful recovery.

Returned to hospital February 7, 1916. Complained of severe pain in right knee. Pain worse at night.

P. I.—About two weeks ago patient had a swelling on the medial side of the right great toe, which was very sore. At the same time the right knee on the lateral side became sore and a little swollen. The sore on toe did not break open but dried up and left a brownish scar. The knee continued to pain and kept the patient awake at night.

Examination shows swelling at outer part of right knee, occupying entire lateral surface and extending about 1 centimetre to the under surface into the popliteal region. Patient was confined to bed and a wet dressing applied to the knee.

February 13th: Pain in knee is less. Temperature varies from 37.2 to 38.4. Blood count: Leucocytes, 18,000; polys, 82 per cent. Patient refuses incision.

February 15. Cast applied to knee.

February 17: Pain worse. Cast removed and compress of Burow's solution applied.

February 18: Pain worse; temperature 37.9 to 38.4. W. B. C., 25,000; polys, 83 per cent.

February 19: Ice bag applied to knee.

February 20: Temperature rose to 39.9. Hot-water bag instead of ice bag. Pain somewhat relieved.

February 21: Patient taken to surgery. Two red inflamed areas were seen on the lateral aspect of the right knee. These were fluctuating and soft. Under local anesthesia the fluctuating areas were opened by incisions and a rubber tube inserted. A considerable quantity of yellow pus escaped.

February 22: Complains of pain on the inside of the knee. Highest temperature, 38.7. Similar nodules and swelling on inside of knee and indistinct fluctuation.

February 23: Abscess on inside of knee opened and drainage tube inserted. Highest temperature, 37.9.

February 24: Feeling much better; slept well; highest temperature, 37.3.

March 10, 1916: Two very small spiculæ of bone directly in skin; one on the right arm near insertion of deltoid muscle in outer groove, the other in popliteal fossa of right leg. These were extirpated, together with the skin containing them, for microscopical examination. Wounds sutured with silkworm gut.

March 15: Sutures removed from arm incision; union per primam.

March 16: Patient removed bandage from arm during night; wound broke open but quite clean. Edges approximated with adhesive.

March 17: Sutures removed from leg incision.

March 20: Hand and arm healing rapidly. Wound in leg clean and healing.

Discharged to come to clinic for treatment.

Small spurs of bone in the abdominal wall projecting with their sharp edges into the skin and giving considerable pain were removed on January 15, April 6, July 30 and December 30, 1917. The muscles and fasciæ in the places ossified by these pieces of bone have entirely disappeared and the lower surface of the bone is adherent to the peritoneum.

X-RAY FINDINGS. *March, 1918*—Hands: Both thumbs and second and third fingers show a diminutive last phalanx.

Abdomen, Pelvis and Right Thigh: The right side of the abdominal wall from the level of the crista ilii downward shows ossification in the muscular strata partly free, partly connecting with the pelvic bones. There is a scoliosis to the right in the lumbar vertebræ (compensatory). The pelvis is tilted from right to left. The right crista ossis ilii stands about 3 cm. higher than the left. The part of the os ilii forming the upper and posterior border of the acetabulum is much broadened and the entire frontal part of the right pelvis is obscured as through a thick veil by the ossification of the overlying soft parts so that the right foramen obturatorium seems obliterated in the picture. The neck of the femur stands in an angle of 160° to the shaft and the head is synostotically fixed in the acetabulum. The large trochanter is imperfectly developed, the smaller one relatively better. The shaft of the bone is less than one-half the thickness of the left femur. The entire anterior space ordinarily occupied by the adductor muscles is here changed to a solid mass of bone, which goes down to slightly above the internal condyle, where it merges with the contours of the femur and tibia in the formation of the completely synostosed knee-joint. The atrophic patella and its ossified ligament occupy their normal positions from the femoral attachment to the insertion at the tuberosity below the much-enlarged head of the tibia.

Right Leg: The outlines of the enlarged tibia can be traced in the picture. The soft parts in the popliteal fossa and all along the posterior part of the leg are nearly entirely ossified. The anterior part and the interosseous space are fairly free. The fibula is comparatively slender.

Right Foot: The foot shows an almost complete synostotic conglomerated mass of the ankle bones and calcaneus, together with the ossified muscles and tendons of the dorsum and part of the planta. There is a faint outline of the cuneiform bones at the base of the metatarsals and in the lateral view a similar one between the astragalus, the scaphoid and the first cuneiform. The metatarsal have retained their separate condition with their part of the interosseous muscles between them. The first metatarsal bone is very much enlarged in circumference through the ossification of its surrounding tissue; the fourth toe is microdactylic through a small metatarsal.

The left hip-joint shows a condition resembling coxa vara.

The left foot shows a partial synostosis of the metatarsal and cuboid bones and the same condition as of the fourth metatarsal as the right.

MICROSCOPIC HISTOLOGY.—Decalcification in trichloracetic acid, stain hæmatoxylin, eosin and Van Gieson. The topographic relations of the various formations of the ossifying process to the epidermis are illustrated in Fig. 9, and present the extreme

distance of the bone formation from the epidermis in any of my sections. To save space and expense, this relation is shown only where essential in the subsequent pictures.

Embedded in the distended and torn meshes of the corium are variously sized nests filled with red blood-corpuscles. Throughout these hemorrhagic masses a few more or less slender tendrils of the torn connecting tissues are remaining. The smaller of these masses show no appreciable changes discernible in the surrounding tissue. In the largest one sees greater tissue destruction within the hemorrhagic deposit and a slightly denser (pressure) approximation of a few connective-tissue fibres along the border encircling the hemorrhage.

There are few connective-tissue fibres within the blood extravasation, they are not so close together, the interstices between them larger, and around the margin of the hemorrhagic depots a circlet of the distended connective-tissue fibres. These fibres are the first indication of the local boundaries in the later ossification.

In these and in many of the later-stage sections a zone of badly stained or totally unstained tissue is observed surrounding the forming ossifications. I claim these to be areas of more or less impaired tissue nutrition caused from pressure of the initial hemorrhage.

In more advanced fields with large hemorrhagic central areas one observes a marked hyperplasia of the immediately surrounding connective tissue. There is a distinct oval shape of the border, forming a growing ring of densely crowded fibres and issuing a communicating network of branch lines throughout the hemorrhagic masses, dividing them into alveolar-shaped compartments. Sometimes these branches do not form and we encounter then in the later stages fully enclosed ossified rings enclosing homogeneous hemorrhagic centers. The progress of osteogenesis is from within outward, the region adjoining the erythrocytes representing the youngest stage. The erythrocytes apparently change into or stimulate the growth of fibroblasts. These cells are forming bone (layers) by apposition, the youngest toward the centre, the older outer ones gradually changing into osseous structure.

In a still more advanced stage, at a period preceding and during the beginning and progression of calcification, the outer encircling ring loses gradually its fibrous structure and becomes a lamellated homogeneous structure, with regular bone corpuscles and canals, or reaches this final osseous stage through an intermediate change from the fibrous to the cartilaginous tissue, in many instances during this metamorphosis the osseous border encloses firmly, and in many parts meets, without any intervening tissue or interior branches, the imprisoned mass of red blood-corpuscles.

This direct chain of transformation, however, is not the only one. In some parts we meet a single row of osteoblasts located nearly exclusively along the inner borders of the bone ring or situated in shallow lacunæ and contributing their share to the bone-formation in the outer ring and along the inner branches. They are very large mononuclear cells, the origin of which remained uncertain, perhaps conjoined transformed fibroblasts, as their gradual structural changes can apparently be followed in successive stages. The cartilaginous route finds its representatives at other places where a condensed connective-tissue mass around a primary hemorrhage changes, with the aid of the enclosed extravasation, from its fibrous condition to a homogeneous undifferentiated hyaline basal structure, with large cartilage cells of principally monocellular type imbedded therein. There the younger inner portion of the ring still shows cartilaginous tissue, while at the same location the adjoining outer part is changing into osseous structure.

This ossifying ring has in many instances an inside lining of single or multiple rows of fibroblasts. These fibroblasts crowd together and lose their identity in joining to build up the homogeneous mass. One sees in places an apparently gradual change of erythrocytes into fibroblasts, the precursors of the bone-tissue, small, spindle-shaped nucleated cells, the youngest not larger than the red blood-corpuscles.

They are lying at the inside of the ring between it and the central mass of unchanged erythrocytes, some, as possible witnesses of their new osteogenetic functions, still recognizable near the growing outer ring of bone tissue. At places there are finger- or fungus-shaped protuberances forming various sized and shaped projections from the outer ring into the hemorrhagic mass. At its inner irregular, ragged edge, blood-corpuscles and fibroblasts are fairly mixed in their genetic endeavor to serve as the building material for the new bone. Osteoblasts also take their part in the neoformation, but, as mentioned before, are nearly exclusively distributed at the inner side of the surrounding bone ring.

Other fibroblasts by crowding closely together form the ground substance in places within the central hemorrhagic mass for the new-bone islets and branches. Some of these are still interspersed with cartilage-like cells, which later, during the process of calcification, give way to the bone corpuscles and their ramifications.

New islets appear in many spots of the central mass. They grow and became confluent, and an intertwining network of new bone structure is formed extending from the inner side of the surrounding ring throughout the inner space. The fibroblasts leading the way for all this new structure extend cobweb-like bands developing into progressive stages of ossifying tissue, new-formed capillaries fill every interstice, and, as a resultant from united efforts, they gradually present the picture of bone and bone-marrow. In another stage the calcified connective-tissue ring around the blood extravasation has consolidated from the inside of the ring outward; the cells have lost their individuality; have formed one solid striated mass with the interspersed bone-cells; calcification and ossification have changed the fibroblasts into an elliptic more or less closed ring of bone tissue surrounding the remaining erythrocytes, which show various stages of apparent transformation. They are more or less closely connected with the inner surface of the bony ring, separated in some places from it by a very slender single layer of spindle-shaped cells; in others, touching it directly.

In these latter places the bone is still in the making, and so closely are the erythrocytes interwoven with the youngest spindle-shaped fibroblasts entering into the new bone-formation, that, contrary to all genetic doctrines, one could imagine the direct transformation of blood-cells into these bone-forming cells, which, in changing from the round- to the spindle-shaped form, become nucleated, growing gradually and less separable form in intermediate stages the ground substance, and, with the appearance of the bone corpuscles, the connecting link between the blood-cells and the finished bone tissue. At other places the above-described cartilaginous route is seen where the inside younger parts of the ring show cartilage tissue, while the adjoining outer ones have already changed to bone substance.

The fate of the central blood extravasation is a varied one. Simultaneously with the apparent participation of the blood-cells in the construction of the bone ring there is an energetic growth of interlacing branches extending from the inner side of the ring, forming a network of slender bands throughout the enclosed space. These branches are at first composed of one layer of spindle-shaped fibroblasts, suggesting their origin from transformed erythrocytes, and inclose in the meshes of the net a proportionate quantity of blood-cells from the original extravasate. Layer after layer is added to the first thickness of the branches, the structure of which with increasing volume, like the outer ring, becomes denser and undergoes the change into bone, forming a progressively continuous network of its own, which still encloses the remnants of the more or less changed erythrocytes. In other places of the sections, occasionally near initial hemorrhages (Fig. 40), some minute specimens of bone are seen, which are solid throughout and the ossification of which has apparently proceeded in adding fibro-osteogenetic layers, filling in the space within the outer border line with a finally solid tissue, thus eliminating the trabecular crosswork, but showing even at these stages in some places the enclosed remnants of the pioneer blood-extravasate, fibroblasts, or less changed erythrocytes.

This apposition from within, however, is not the only means of osseous growth. In a few places there also is a small aggregation of fibroblasts at the outside of the ossifying ring adding to its growth, but no osteoblasts appeared anywhere on the outside, showing that their location seems strictly limited to the inner aggregation of erythrocytes. In some sections a hypertrophic growth of outer connective tissue, participating in the ossifying process, may be seen to enter through a gap in the branches of the partly open osseous ring and closing it first with a connective-tissue bridge, which later ossifies, having previously sent a continuation of the ingrowing tissue from the gap toward the inner side of the ring. In others there is a predominant erythrocyte participation in the bone-formation of the ring.

We may even in some isolated instances get pictures that suggest a prevailing bone-formation from the outer side of the ring, the connective tissue changing directly to bone-tissue by calcification and development of bone-corpuscles and canals.

In these cases the inner erythrocytes play a minor part in the final ossification; the major part is taken by the connective tissue direct.

The structural changes of the hemorrhagic nucleus do not always keep pace with the development of the bony ring in all parts of an ossified nodule nor in every specimen of a series, and allowances have to be made for more or less of the erythrocyte-filling dropping out during the course of preparation, especially from paraffin sections. Thus we see pictures where a completely unbroken osseous wall closely surrounds a structurally, nearly unchanged, inner hemorrhagic mass. In others a more active participation of the connective-tissue growth in osteogenetic activity and a rather regressive metamorphosis of the enclosed hemorrhagic centre part.

In all these various formations the primary incentive is the hemorrhage with a consecutive connective-tissue growth, and whether the final result is a spongeoid or a solid bone, the route to it directly from the erythrocytes into ossifying fibroblasts or from those via fibrous connective tissue, hyaline or fibrous cartilage, with or without the help of osteoblasts, we have always to look back to the first hemorrhage as the starting point.

Analytical Study of Reported Cases

Before we proceed to analyze these 120 cases, we must explain that five of the number (four male and one female) were only entered in the list on account of their having figured in all the former compilations, and might be classified as "overlooked" by later investigators. They are the cases of E. Schwarz (No. 35), Kronecker (No. 36), E. Lexer (No. 50), Eichhorst (No. 53), and Krause and Trappe (No. 86). Neither of these is myositis ossificans progressiva, and they must be deducted from the total combined number (120-5-115). Four of these are males, one a female, so we have among 115 true cases 68 males, 42 females and 5 where the sex has not been recorded—a somewhat different proportion from former statistics. (Rolleston, 5 to 1; Nissim and Weill, 38 to 12.)

Their ages at the time when the first symptoms were observed were in the period before Muenchmeyer's classification (1868):

Males—0-5, 2; 5-10, 0; 10-15, 3; 17, 1; 18, 2; 21, 1. Females—8, 2; 5, 1; not recorded, 8. Not myos. prog., 5.

After Muenchmeyer's: Males—0-2, 19; 2-5, 23; 5-10, 7; 13, 2; 14, 1; 18, 1; 19, 1; ?, 1. Females—0-2, 10; 2-5, 17; 5-10, 10; 12, 1; 20, 1. Sex and history unknown, 1.

FIG. 1.—*a*, area of ossification in abdominal wall; *b*, central hernia.

R. FIG. 2. L.

FIG. 3.

FIG. 4.

FIG. 5.

FIG. 6.

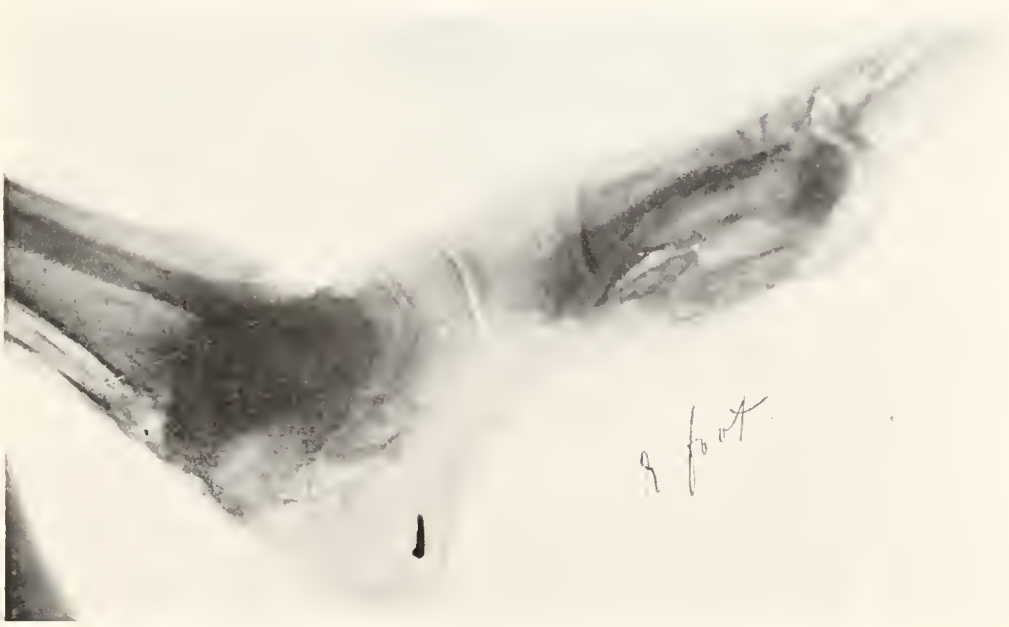


FIG. 7.

Fig. 8.

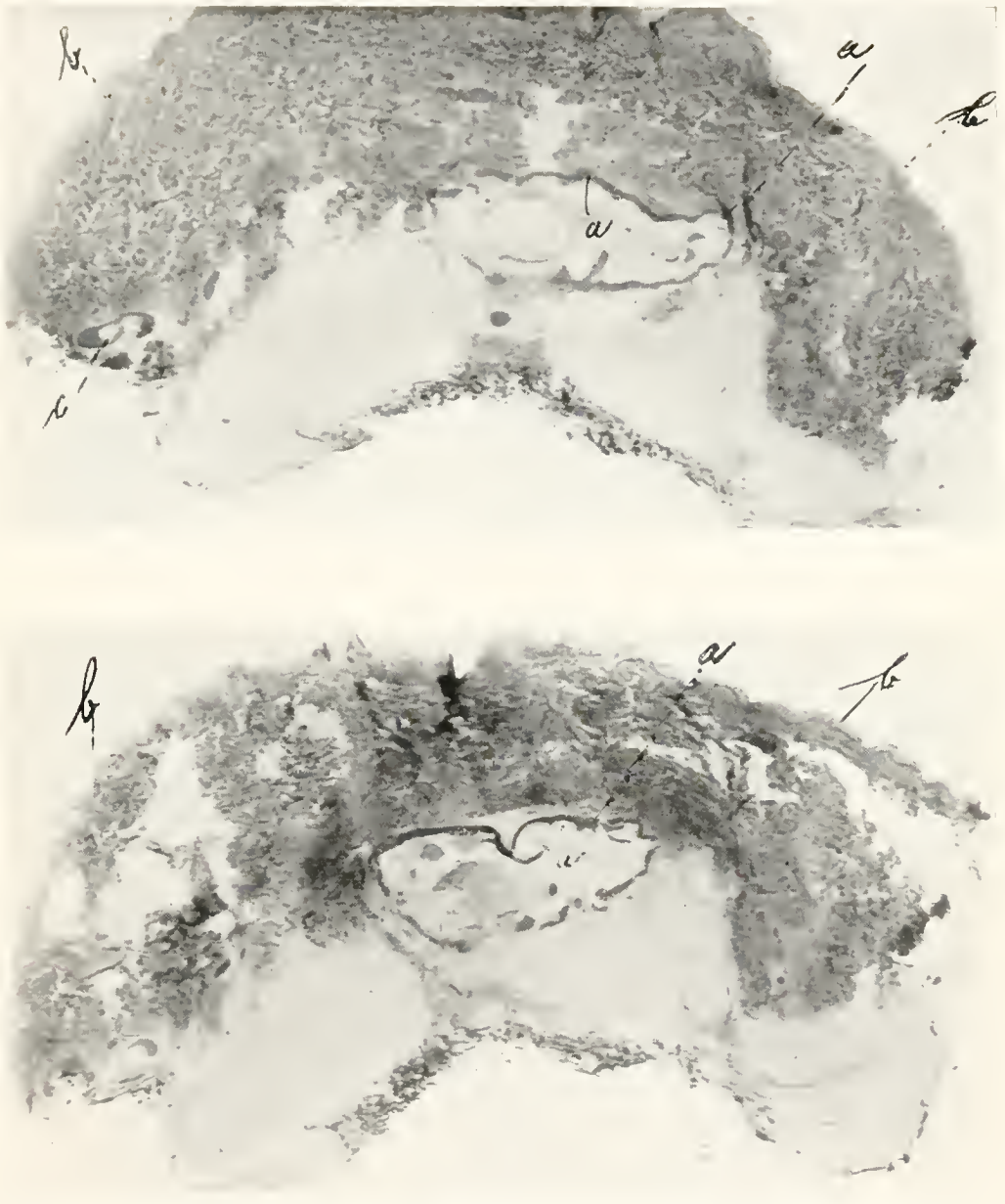


FIG. 9.—Topographic view of skin sections with bone ring. *a*, bone ring; *b*, epidermis.

A

B

C

FIG. 10.—The initial hemorrhage. *A*, low power (oc. II, obj. 16). *a*, hemorrhage (earliest stage); *b*, invasion of surrounding area. *B*, low power (oc. II, obj. a *). *a*, hemorrhage; *b*, epidermis. *C*, higher power (oc. II, obj. 4 ap). *a*, hemorrhage; *b*, connective tissue.

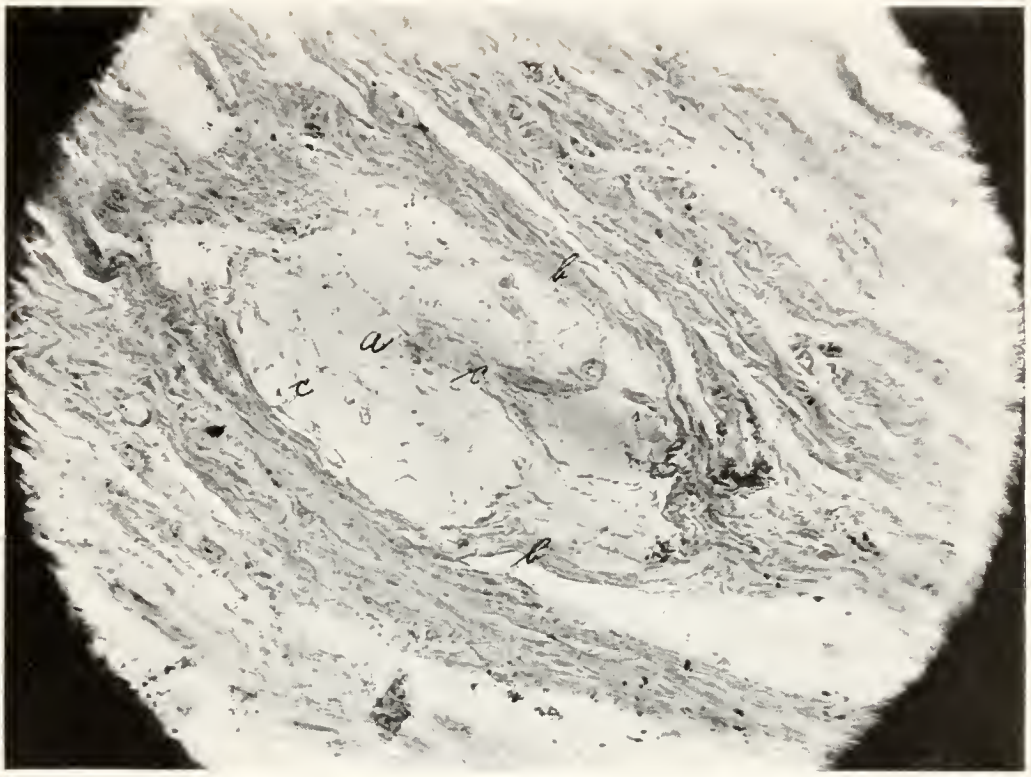
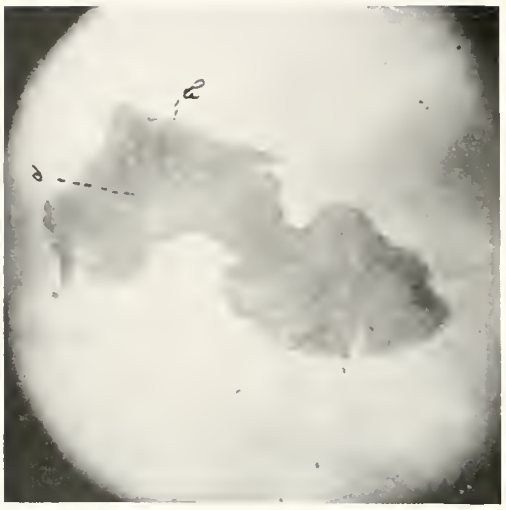


FIG. 11.—*a*, primary hemorrhage; *b*, beginning of boundary formation; *c*, displaced and torn shreds of connective tissue strands. Enlargement of Fig. 10 C.

FIG. 12.—*a*, primary hemorrhage; *b*, invasion into surrounding normal tissue. Enlargement of Fig. 10 A.

A



B

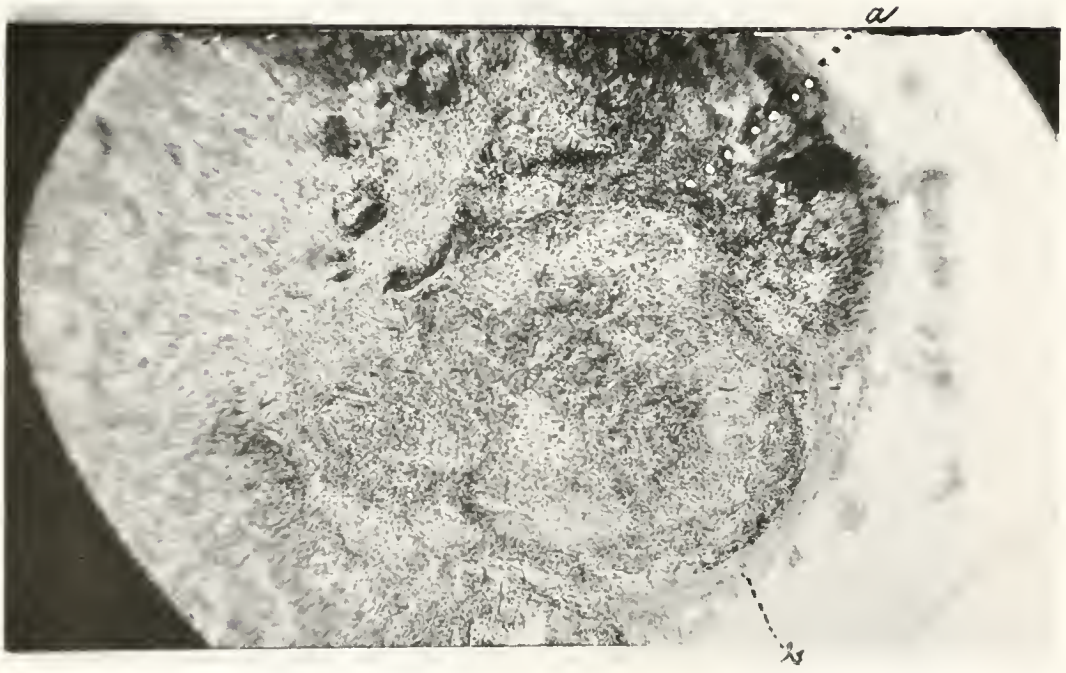


FIG. 13.—Initial hemorrhage with beginning encapsulation. *A*, low power (oc. II, obj. a *). *a*, hemorrhage; *b*, epidermis. *B*, higher power (oc. II, obj. 8). *a*, hemorrhage; *b*, beginning connective-tissue circlet.

A

B

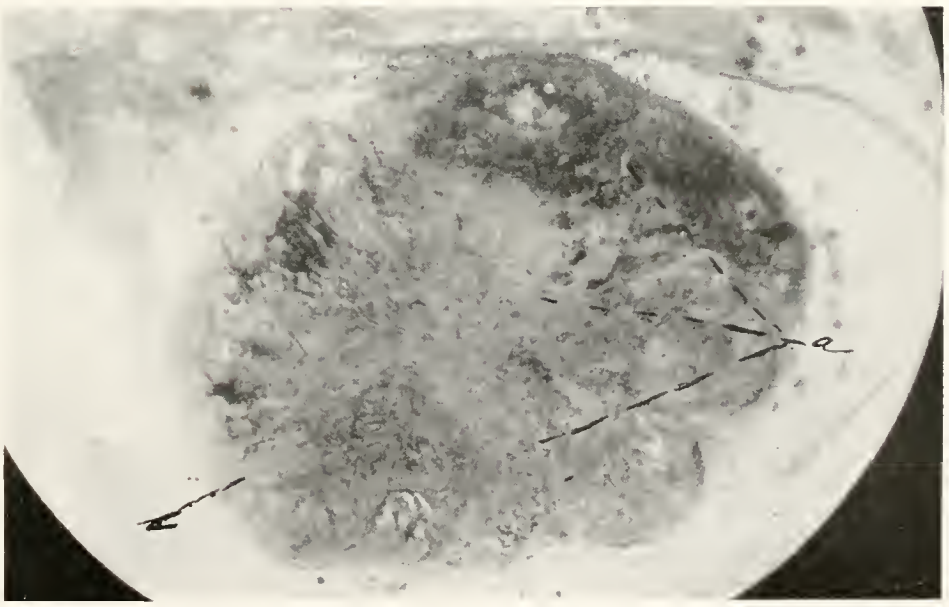
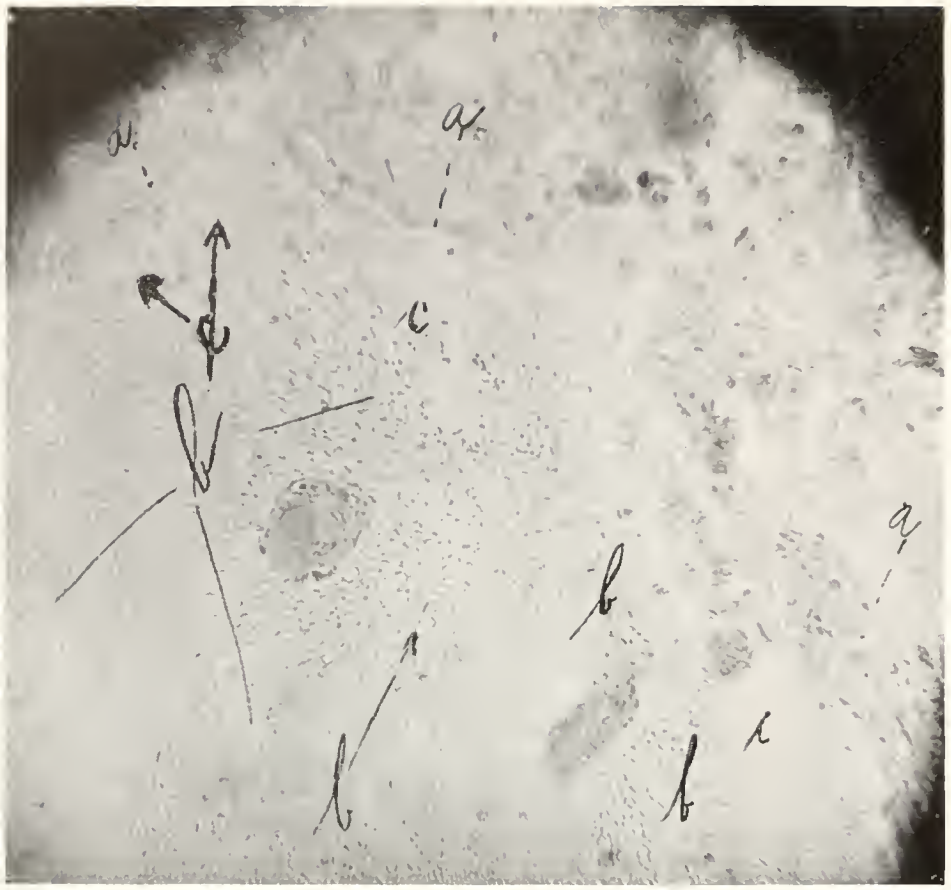


FIG. 14.—Initial hemorrhage with advanced connective-tissue encapsulation. *A* (oc. II, obj. 6). *a*, hemorrhage; *b*, connective-tissue circlet. *B* (oc. 5, obj. 8). *a*, hemorrhage; *b*, connective-tissue circlet.

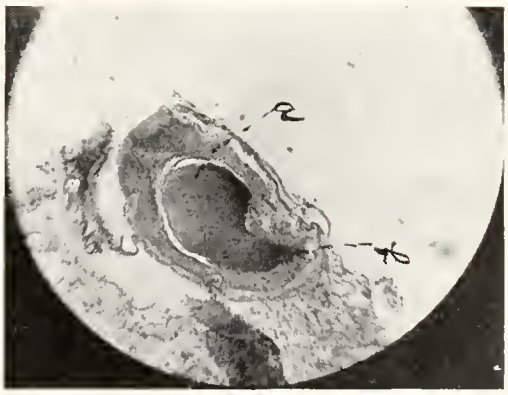
A



B

FIG. 15—*A*. *a*, beginning of ring formation; *b*, erythrocytes; *c*, change of form of erythrocytes to spindle shape. *B*. Ring formation nearly completed. *a*, newly formed ring; *b*, erythrocytes; *c*, slight projections from ring into erythrocyte mass.

A



B

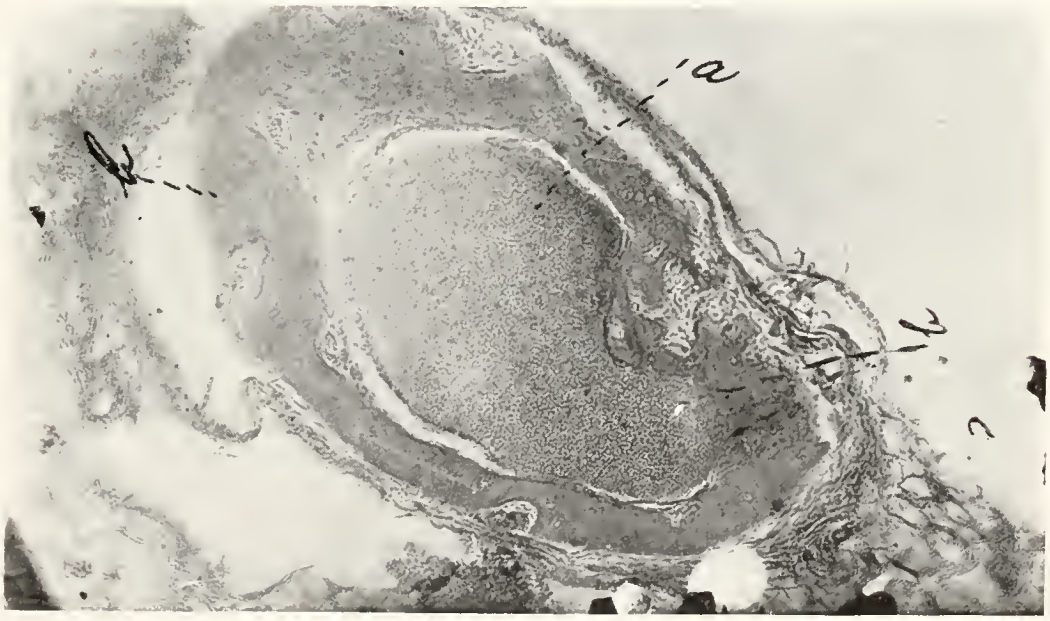


FIG. 16.—Solidification of outer connective-tissue ring, hemorrhagic centre. *A*, *a*, hemorrhage; *b*, solidified osseous ring (oc. II, obj. 16). *B*, *a*, hemorrhage; *b*, solidified osseous ring (oc. II, obj. 8).

A

B

FIG. 17.—*A*. Solid hemorrhagic mass in close apposition to ring in advanced stage of ossification (enlarged part of Fig. 17). *a*, bone ring; *b*, mass of erythrocytes; *c*, change of form of erythrocytes to spindle shape. Enlargement from part of Fig. 16. *B*. Solid hemorrhagic mass in close apposition to ring in advanced stage of ossification (enlarged part of similar section as Fig. 16). *a*, bone ring; *b*, mass of erythrocytes; *c*, change of form of erythrocytes to spindle shape.

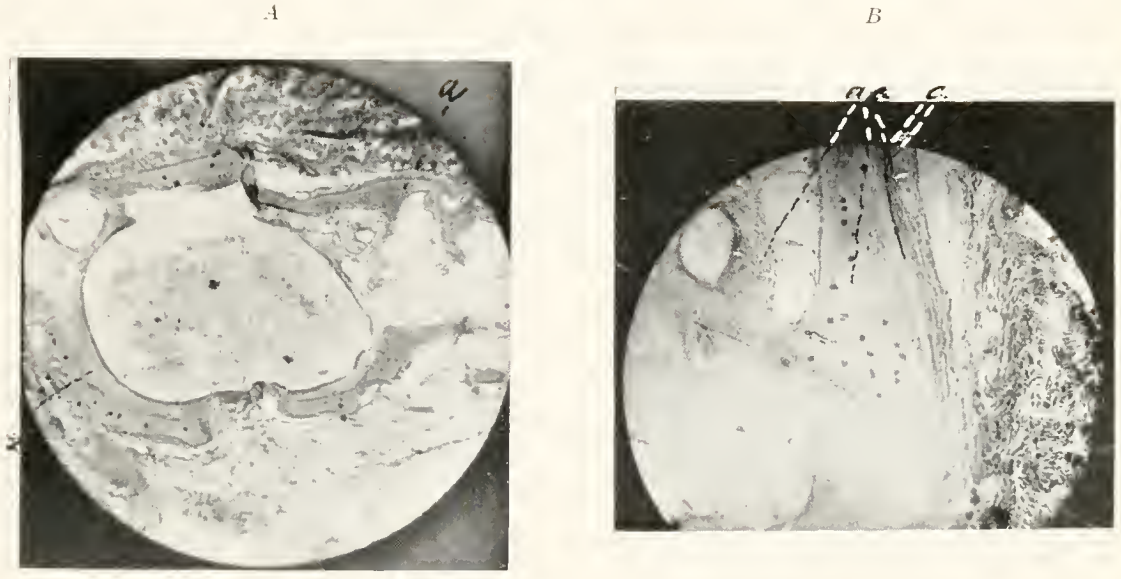


FIG. 18.—Cartilaginous tissue bordered on inside of ring by fibrous on outside by osseous tissue. *A*, *a*, place where magnified picture was taken (oc. 2, obj. 16). *B*, *a*, fibrous part; *b*, cartilaginous part; *b'*, fibroblasts changing into cartilage cells; *c*, osseous part (oc. 6, obj. 8). Enlargement of *A*, (*a*).

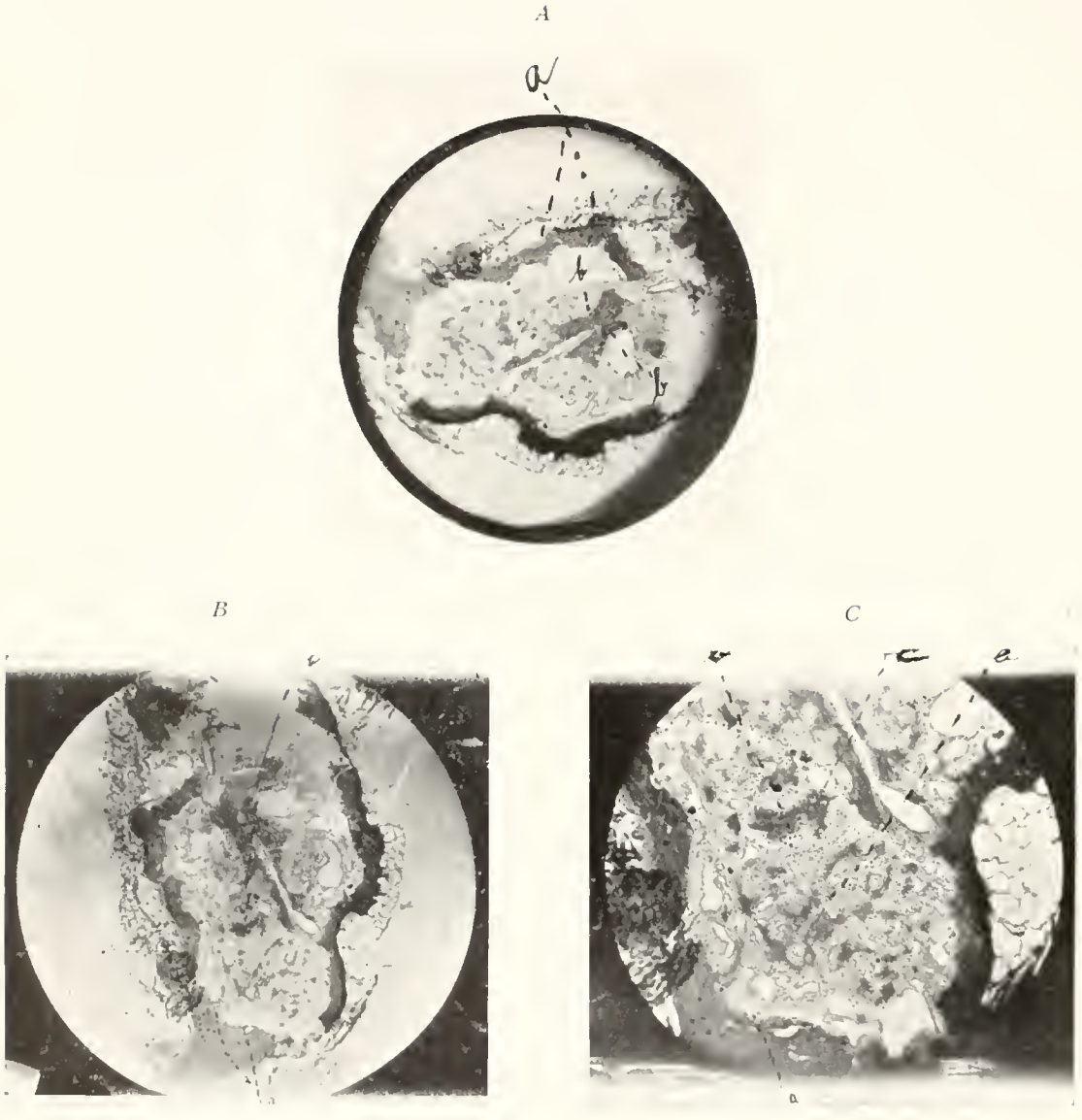


FIG. 19.—*A*. Osteoblasts on both sides of ossifying branches. *a*, outer ring; *b*, osteoblasts (oc. —,obj.—.). Osteoblasts on inside of ossifying ring and erythrocytes participation. *B*. *a*, osteoblasts inside ring; *b*, erythrocytes participation and apparent change (oc. II, obj. 8). *C*. *a*, osteoblasts inside ring; *b*, erythrocyte participation and apparent change; *c*, osteoblasts at branches.

A glance at these two groups shows the first observations of the initial symptoms at a remarkably earlier average in the second group than in the first. Although the number of the first is too small for a valid comparison, still the influence of Muenchmeyer's publication in drawing the attention of the profession to this mysterious disease cannot be ignored.

Since Helferich's enlistment of the congenital digital deformities among the symptoms of this affection their registration has not been followed by all authors. We believe that they are present in practically all genuine cases, and consequently of the ninety-three cases of this series the thirty-one where they have not been registered demand an explanation. In the first place, we have to deduct the five spurious cases already mentioned, and one of the other remaining (Bernacchi, No. 42) shows plainly on the accompanying photograph microdactylia of the thumbs. All of the other twenty-five who answered my inquiries (six) as to the presence or absence of these congenital deformities in their cases admitted that they had failed to look for them. In fact, all the thirty-one histories, with one exception, contain no reference to this symptom, and in my abstract of their cases I always inserted under the heading of congenital deformities, "not mentioned." There is only one author (Salvetti, No. 79) who specially dwelt *on the absence* of microdactylia of either fingers or toes in the description of his case, but his subject was very young (four years) at the time of observation, when slight differences in the size of fingers or toes may easily escape notice, and a decisive X-ray showing the condition of the epiphyses was not taken. Several of the remaining cases have as their only records brief demonstrations before medical societies. My conviction, therefore, that every genuine case of myositis ossificans progressiva shows mycrodactylia of either fingers or toes, or both, holds good in spite of the nineteen questionable cases. Speaking loudly in my belief in the failure of noticing this symptom stands the example of the great master of close observation, Rudolf Virchow, who failed to mention it in the history and description of his celebrated case (No. 46), while attention to the presence of microdactylia was called later by Gerhard in the discussion following the demonstration in the Berlin Medical Society. The malformations present in the cases were distributed as follows:

Cases after Helferich (1879): Thumbs and big toes and little fingers, 6; all fingers and big toes, 2; thumbs and fourth fingers, 1; thumbs and big toes, 24; big toes, 29; fourth and fifth toes, 1.

The very marked preponderance in this compilation of the big toes, they being alone or in combination with other terminal parts of the extremities the seat of the brachydactylic deformity, is shown by their being represented sixty-one times in the sixty-three available cases.

One does not wonder that a disease, so baffling in its course, from the first causes to its ultimate state, should invite the speculative as well as the patiently investigating observer to lift the obscuring veil and solve this embarrassing puzzle.

One of the first attempts to explain the mysterious process was the

theory of a retention of calcium in the organism due to a faulty metabolism which failed to excrete its quota of lime. This excess of lime in the fluids and the tissues was presumed, by its mechanical or chemical action, to exert an irritating influence and cause ossification.

H. Duvy, in Abernethy's case, was the first to examine the urine of these patients for phosphate of lime and found the quantity lower than the average. This was confirmed by Dittmeyer[1], Helferich[2], Pinter[3] and Partsch[4]. Duvy, Partsch and Pinter found in addition the excretion of urea normal; of uric acid, kreatinine and earth phosphates, diminished.

As all these investigations were confined to the urine alone, without regard to fæces or to exact account of the intake, they are valueless and must be rejected.

Very exact and complete analyses, however, have been made by Burgerhout[5] and Austin[6], both giving absolutely negative results. As Burgerhout's excellent work, besides being buried in a dissertation, is written in Hollandish, and Austin's has been published separately from Painter and Clarke's case, which served for the investigation, I shall give both their tables here in full. They are self-explanatory and Burgerhout's deductions are conclusive.

Burgerhout's comments on these tables are as follows: "According to the results of the examinations of our patients, no difference in the exchange of matter seems to be observable. The metabolism according to the figures certainly does not deviate from that of normal persons. Under ordinary circumstances the resorption of the necessary amounts of nourishment leaves nothing to desire. The greater increase in this patient's bone-tissue produces no typical alteration in his metabolism through the composition of food which at all times exercises a governing influence upon the phosphates

The daily diet during A and C was: 1.5 l. milk; 250 gr. bread; 100 gr. rice; 75 gr. butter; 50 gr. sugar, and one cup of Liebig's bouillon, calculated as follows:

	Albumen	Fat	Carb. Hydrates
1.5 l. milk	46.875 gr.	45 gr.	67.5 gr.
		2.5 gr.	150. gr.
250 gr. bread	21.875 gr.		
100 gr. rice	6.875 gr.	0.9 gr.	77.5 gr.
75 gr. butter	0.468 gr.	65.25 gr.	.....
50 gr. sugar	.....	.....	50 gr.
	76.2 gr.	113.5 gr.	345 gr.

Equal to 2782 calories, or about 32 calories per kgr. bodyweight. (Patient lying nearly motionless in bed.)

[1] Op. citat.
[2] Op. citat.
[3] Op. citat.
[4] Op. citat.
[5] Op. citat.
[6] *Journal of Medical Research*, 1907, vol. xvi, p. 452.

TABLE A

FIRST PERIOD URINE ANALYSIS

Date	Quantity	Sep W	Cl	P_2O_5	N	CaO	MgO	Kr	Body Weight
June 29....	1280	1016	4.268	1.805	8.996	0.228	0.076	0.177	68.7 kgr.
June 30....	800	1032	4.557	2.292	12.611	0.197	0.074	0.532	
July 1.....	560	1029	3.971	1.053	6.372	0.207	0.057	0.181	
July 2.....	1430	1020	6.759	1.837	9.810	0.262	0.073	0.220	
July 3.....	1180	1023	5.792	2.136	9.416	0.244	0.085	0.120	
July 4.....	1020	1023	5.192	2.025	6.569	0.235	0.091	0.451	68.3 kgr.
			30.539	11.148	53.774	1.373	0.456		
Analysis of fæces during entire period.....			0.047	15.814	4.291	11.369	0.627		
Total urine and fæces.....			30.586	26.926	58.065	12.742	1.083		

TABLE B

Hunger period: nothing taken but water, of which 4.1 l. was consumed.

URINE ANALYSIS

Date	Quantity	Spec. W	Cl	P_2O_5	N	CaO	MgO	Kr	Body Weight
July 5.....	2100	1012	6.108	1.785	8.526	0.179	0.074	0.130	68.3 kgr.
July 6.....	1210	1016	1.219	1.954	7.996	0.196	0.102	0.416	
July 7.....	1730	1014	1.677	2.197	8.910	0.398	0.106	0.294	65 kgr.
			9.004	5.936	25.432	0.773	0.282		
Analysis of fæces during entire period.....			...	4.689	1.856	3.218	0.188		
Total, urine and fæces.....			9.004	10.625	27.288	3.991	0.470		

TABLE C

LAST PERIOD URINE ANALYSIS.

Date	Quantity	Spec. W	Cl	P_2O_5	N	CaO	MgO	Kr	Body Weight
July 8.....	710	1026	2.237	1.260	8.300	0.402	0.086	0.499	65 kgr.
July 9.....	500	1030	2.729	1.205	8.176	0.266	0.062	0.678	
July 10.....	530	1031	3.469	1.102	7.984	0.241	0.044	0.521	.....
July 11.....	710	1019	5.206	1.441	9.164	0.258	0.048	0.481	
July 12.....	1070	1018	5.058	1.198	6.861	0.189	0.039	0.284	
July 13.....	1320	1018	5.279	2.086	9.203	0.211	0.059	0.437	67.3 kgr.
			23.978	8.292	49.788	1.567	0.338		
Analysis of fæces during entire period.....			0.056	11.808	2.956	8.296	0.529		
Total, urine and fæces.....			24.034	20.100	52.744	9.863	0.837	.....	

TABLE D

	Cl	P_2O_5	N	CaO	MgO
Total hunger period.....	9.004	10.625	27.288	3.991	0.470.
Deduct 4.1 water l. consumed.....	0.500	....	....	0.463	0.023
Real loss during hunger period.....	8.504	10.625	27.288	3.528	0.447

TABLE E

	Cl	P_2O_5	N	CaO	MgO
Total first period	30.586	29.962	58.065	12.742	1.083
Total third period	24.034	20.100	52.744	9.863	0.873
Difference	6.552	6.862	5.321	2.879	0.246
1.3 l. more water consumed during 1st period than in 3rd	0.159	....	....	0.147	0.017
Real retention in 3rd period	6.393	6.862	5.321	2.732	0.229

AUSTIN'S TABLES[7]

	Daily CaO in food	Daily CaO in fluid	Daily Total intake	Daily CaO in urine	Daily CaO in fæces	Daily Total output	Daily Balance
First period November 28. Average of two series of seven days each	.880 gr.	.366 gr.	1.246 gr.	.472 gr.	.787 gr.	1.257 gr.	.011 gr.
3rd series	.560 gr.	.012 gr.	.572 gr.	.425 gr.	.473 gr.	.899 gr.	.327 gr.

Substance taken poor in calcium: Roast beef, white bread, grapes, butter, oranges, chicken, spring water.

and alkalines whereupon it hinges in these cases. During the hunger-period, when this disturbing influence ceases, we see distinctly that this bone-tissue, by its increased quantity, comes more into the foreground, and how our patient, in this way living at the expense of his body-tissues, also uses more of his increased amount of bone-tissue than normal persons." Austin's exact tables show a similar result.

These tables, representing a complete analysis of the entire intake and the total output through urine and fæces during three series of seven days each, show absolutely normal conditions and no retention of lime.

With the multiplicity of cases and with modern methods the pathological anatomy of progressive ossifying myositis has been fairly well cleared. No longer prevails the old definition of Virchow[8], who classified the pathological products as belonging under the heading of multiple osteomata and that the swellings representing such distinctive tumors always originate from the periosteum.

This view had also been expressed by Eberth[9] subsequent to his findings at an autopsy of a case of myositis ossificans progressiva which showed the ends of the muscles and their tendinous insertions ossified, the bodies of the muscles though relatively intact. This opinion was, moreover, shared at a much later date by Pincus,[10] who denied to our malady the distinction of being a type *sui generis*, adding that it belonged by all means to the class of osteomata and exostoses and possessed the indubitable characteristics of a tumor-disease, its original starting-point being

[7] A. E. Austin: Calcium Metabolism in a case of Myositis Ossificans; *Journal of Medical Research*, vol. xvi, March to July, 1907, p. 451.

[8] Die krankhaften Geschwuelste and op. *citat.*

[9] Ueber multiple Knochengeschwuelste, Deutsche Klinik, 1862, No. 9, p. 91.

[10] Op. citat., 1897.

the periosteum, and its casual element the hemorrhage from traumatism during birth.

Mays[11] was inclined to compromise. He asserts the pathological process to be in the osseous system along the course of tendons and fasciæ and in the intramuscular connective tissues as well as in the loose connective tissue between the muscles; but he affirms at the same time that the entire tissue complex is not subjected to this change, this anomaly being nowhere encountered in the subcutaneous tissue, affecting only the connective tissue of that apparatus which Virchow has singled out as a seat of the multiple osteomata, "the apparatus of motion."

He was disposed to range this disease with the exostoses, although his microscopical examinations point toward an adoption of the theory of a congenital predisposition to the ossification of the intramuscular connective tissue, the fasciæ and aponeuroses.

Muenchmeyer[12] was the first to pronounce this view, and in separating it from all cases of multiple osteomata he drew a concise picture of the malady and characterized it as a peculiar and distinctive constitutional anomaly.

Cahen[13] pronounced, as the first, an absolute identity in the microscopical pictures of traumatic and progressive ossifying myositis and a direct causal connection between the hypertrophy or the intermuscular connective tissue and ossification. Other reliable microscopical observers confirmed, in more or less complete form, these findings.

Lexer[14] asserts that the excessive increase of the intramuscular and connective tissue-cells and the formation of an indifferent tissue, rich in cells and profusely vascularized appearing with a simultaneous atrophy of the muscular tissue, is the forerunner of ossification. These new connective-tissue cells gradually assume large round forms and then change into cartilage cells.

Kraske[15] believes that those osseous neoformations, not emanating directly from the bones, are ossifications extending from the insertion of the muscles into the tendon-fibres and fasciæ and the connective tissue of the muscles. The muscles do not participat in this change, and waste away through pressure-atrophy and fatty degeneration.

In Goto's[16] very extensive microscopical studies of his case the author arrives at the conclusion that the primary process is the hyperplasia in the fascia or aponeurosis, tendon or periosteum, also, under certain circumstances in the ligamentary apparatus.

The muscular interstitial tissue, which most authors describe as the primary seat of the affection, is, according to his investigations, only secondarily invaded from the neighborhood. He would call the disease logically hyperplasia fascialis ossificans progressiva. He divides the course of the disease into three stages:

1. The stage of connective-tissue hypertrophy without inflammation.
2. The stage of fibrous induration.
3. The stage of ossification.

The new-formed bone always shows a normal bone structure.

In this paper Goto attaches no importance to the hemorrhagic extravasations found in all of his specimens excised at an early stage of the local affection. Pieces excised later still showed traces of an earlier hemorrhage, but mostly absorbed. He expresses the belief that the hypertrophy of the connective tissue takes place so rapidly and energetically that the muscular elements in spite of their elasticity have no time

[11] Op. citat.
[12] Op. citat.
[13] Op. citat.
[14] Op. citat.
[15] Op. citat.
[16] Op. citat.

to escape the process of distention. They, together with the thin-walled blood-vessels, are finally torn, and thus cause the secondary hemorrhagic foci. We will see later that his experimental work for traumatic ossifying myositis shows another aspect of the importance of blood and blood-vessels for the ossifying process.

The microscopical findings in my own case, while confirming that the primary tissue change takes place in the extra and intramuscular connective tissue, add to this a hitherto unknown feature of this disease, a bone-formation in the skin—a feature contrary to all previous experience. It involves principally the corium and therefore does not definitely trespass into the ectoderm; but its independent and isolated occurrence, in even the connective-tissue layer of the skin, has not been observed before. This find more than any other proves the primary tissue affected in this malady to be the connective tissue of the mesoderm, and the secondary and passive part, the muscular tissue. The significant presence of hemorrhagic extravasations as forerunners and foci of the new-formed cartilage and bone will be considered in connection with the question of the immediate cause for the bone-formation.

In considering the various causes theoretically or experimentally advanced for the explanation of bone-formation in this disease, I shall also include those for the *traumatic* myositis ossificans. While those based on the trauma itself can be easily excluded, we must recognize that the various stages in the microscopical pictures of the process of ossification are identical in both and may be profitably examined and reviewed in common.

The authors writing on this subject seem, with few exceptions, to be divided into two camps, one ascribing the stimulating cause to the presence of lime deposits, the other to blood extravasation into the tissues, due to the rupture of thin-walled new-formed blood-vessels. There is lacking in both explanations the ulterior reason or proof for the presence of lime or of the free blood, the theory of metabolic lime retention being untenable in the face of the exact analyses of Burgerhout and Austin, and except in the cases of traumatic ossification none either for the extravascular blood.

After a series of experiments and histological work, Berlies[17] arrives at the conclusion that the muscular osteoma (rider's bone) is not a tumor, but is formed in connection with the tearing off of a flap of periosteum. This is the basis of the new formation of bone, the detached flap surrounded by the muscular fibres acting as a bone-graft within the body of the muscles.

A development of bone tissue may also be gained by direct transformation of cartilaginous embryonic connective tissue, or of fibrous connective tissue.

Rathcke[18] communicates an interesting case bearing upon this latter phase of development. A sailor 28 years old was hurt by the fall of a sailing yard on his right thigh. A swelling resulted immediately afterward, which gradually hardened.

[17] Etude histologique et expérimentale des Ostéomes musculaires. Arch. de méd. expériment. Paris, 1894. Tome 6, p. 600.

[18] Ueber die Ursache d. gelegentl. Auftretens v. Knorpel bei d. Myosit. ossif. Arch. f. Entwicklgsmechanik d. Organismen. Bd. vii, 1898, p. 398.

Extirpation some time later of this hardened mass showed an intersection of the vastus internus, with bands of interstitial connective tissue growing harder the nearer it approached the bone. In the peripheral parts the growth was a cartilaginous tissue, but near the bone it was osseous and had to be separated with a chisel. Rathcke gives the explanation that where free motion exists, the tissue has to yield to changes of location and friction of the various tissue layers among each other and the cartilage adapts itself to these conditions, whilst the nearer the bone the traction and pulling is more and more prevented by its immovability, and only those cells survive which can offer the necessary resistance by their osseous transformation. This illustrates also the theory of Roux.[19]

Cornil and Condray[20] grafted entirely detached pieces of periosteum into the leg muscles of young dogs and produced thereby growth of osteomata in these muscles. The microscopical study of these and of extirpated human myosteomata convinced them that the bony tissue of these tumors grows at the expense of the connective tissue and not at that of the more or less transformed muscular tissue.

John Morley's[21] experiments on rabbits by scraping the periosteum and crushing the overlying muscle, produced bone-growth into the muscles. His specimen and X-ray pictures show this new growth to emanate exclusively from, and remain in direct continuation with, bone, forming an exostosis extending into the muscular tissue.

Another theory that some component of the joints, evidently the synovia, plays an important part in the ossification of myositis ossificans traumatica, was advanced by Ewald.[22] The lesion produces a tear in the capsule, some of the synovia escapes and then exercises the peculiar stimulus leading to ossification.

The experiments of Fabris[23] were based upon similar suppositions. His observation was that most cases of myositis ossificans traumatica occur in the neighborhood of articulations. He injected synovia into the muscles of dogs, but his results were absolutely negative; not the slightest trace of osseous tissue could be found in the injected area.

Among those who advocate that the presence of lime is the important cause of ossification, Cohn[24] believes that in a fair number of cases a chronic mechanical irritation by lime alone may lead to ossification (in blood-vessels).

Marchand[25] asserts that genuine bone may be produced by connective tissue everywhere where sufficient lime salts are available, *i.e.*, where calcification has preceded. Without the presence of lime no bone could be found, in spite of the most energetic hyperplasia of connective tissue.

Rohmer[26] believes that the first condition for the production of bone is the new formation of a highly vascularized connective tissue, and the second the presence of lime.

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[19] Gesammelte Abhdlgn. Ueber Entwickelgsmechanic, Bd. ii, No. XVIII. Leipzig, 1895.

[20] Sur le mode de dévelopement des ostéomes musculaires d'origine traumat. Bulletin de l'Académie de méd. Paris, 1907. Pp. 597–600.

[21] Traumatic intramuscular ossification. *British Medical Journal,* Dec. 5, 1913, p. 1475.

[22] Zur Aetioligie d. Myos. ossif. traumat. Zentralbl. f. Chir. 1910, No. 22, p. 771.

[23] Fabris: Sulla genesi della miosite ossificante. *Gazetta degli Ospedali e della Cliniche,* 1911, No. 42, p. 443.

[24] Cohn: Ueber Knochenbildg. in d. Arterien. Virch. Arch. Bd. 106, 1886.

[25] Marchand: Verhdlgn. d. deutsch. Pathol. Ges. Ber. ueb. d. 2 Tag. Aachen 17-20, September, 1900.

[26] Ueber Knochenbildgn in verkalkten endocarditischen und endarter. Heerdez Virch. Arch., Bd. 166, p. 13.

A similar view as to lime has been entertained by Pollak[27], who observed cases of heterotopic bone-formation in the lungs. It always occurred around calcified necrotic parts. His case (No. 21) showed an additional ossification of the stylohyoid; case No. 24, exostoses on the lumbar vertebræ.

Roepke[28] joins in the conclusions that the presence of lime salts within a traumatically inflamed connective tissue furnishes the irritation and cause for bone-formation.

Georg B. V. Gruber[29] emphasizes again the absolute identity in the microscopical pictures of slides of traumatic myositis ossificans circumscripta and myositis ossificans progressiva, and adds that wherever young and very vascular connective tissue meets with previous lime deposits an osseous formation is possible without any relation to periosteum. Such has been observed in blood-vessels, lymphatics, glands, brain, liver, kidneys, vas deferens, etc. He also mentions the possibility of secondarily encysted blood or lymph deposits being the source of lime for bone-formation. In a previous paper[30] the same author arrived, among others, at the following conclusions: "That the periosteal genesis in myositis ossificans traumatica is improbable and is absolutely excluded except in cases where the muscle-bone is in direct communication with the skeletal bone; and that the ossifications of the muscles are caused by the united action of young cellular connective tissue meeting the deposits of calcium, or of being stimulated to ossification by its presence in the tissues.

"In order to decide the problem of the genesis of muscular ossification or to solve that of heterotopic ossification in general, the questions regarding the calcium metabolism and the calcification of tissues must be answered. Until now the acceptance of a certain individual disposition to such ossification cannot be dispensed with entirely."

Dibbelt[31] drew attention to the difference in the forming of bone between carnivorous and herbivorous animals, dogs instancing the difficulty among the carnivorous, rabbits the case of the herbivorous. He attributed these diversities to the high calcium metabolism of the herbivores, and supported this theory by animal experimentation. He fed rabbits on food poor in calcium and caused the formation of fibrous tissue in place of bone, the addition of calcium to the food effecting a metaplastic change of tissue into bone.

That free blood acts as a factor in the bone-producing process has been very frequently and persistently claimed by many authors.

Cahen[32] claims this in his paper where he asserts as the first, that the microscopical picture of myositis ossificans traumatica is identical with that of progressive. In his case the hemorrhage caused by the trauma marked the place of origin of the later ossification.

E. Cordillat[33] reports the case of a young soldier, 21 years, entering a cavalry regiment, who, between November 24, 1894, and January 19, 1895, rode altogether about fifteen miles, and who presented himself on the latter date with an ecchymosis, the size of a dollar, on the left leg posteriorly, and a hard, bony mass at the inside of the femoral vessels occupying the entire middle adductor, a length of 17 cm., with

[27] Ueber Knochenbildgn. in der Lunge. Virch. Arch., Bd. 165, p. 129.

[28] Z. Kenntn. d. Myositis ossif. traumat. Arch f. klin. Chir., Bd. 82, p. 81.

[29] Weitere Beitraege z. pathol. Anat. d. umschrieb. Muskelverkng. nebst Bemerkgn. z. Myositis ossif. ueberhpt. Mitth. a. d. Grenzgb. d. Med. and Chir., 1914, Bd. 27, pp. 762-786.

[30] Ueber Histol. & Pathogen. d. circumscript. Muskelverkng. Jena, G. Fischer, 1913.

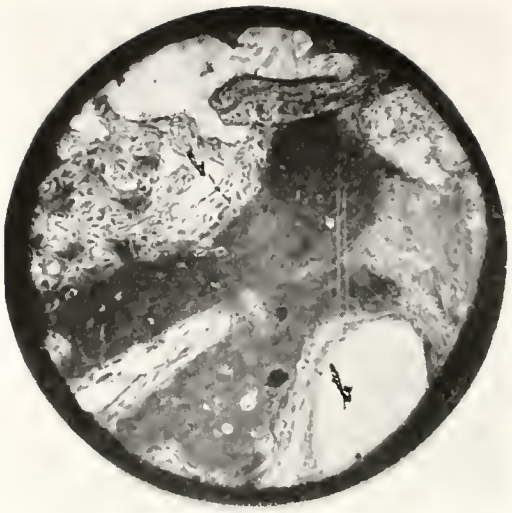
[31] Beitraege z. Histogenese d. Skelettgewebe & ihrer Stoerungen. Beitraege z. pathol. Anat. & z. allgem. Pathologie, 1910. Bd. 48, p. 147.

[32] Ueber Myos. ossif. Deutsche Zeitschr. f. Chir., Bd. 31, 1890, p. 372.

[33] Ostéome bilatéral du moyen adducteur. Arch. d. Méd. et de Pharmac. militaires, 1895. Tome, 26, p. 427.

FIG. 20.—Osteoblasts participating in bone formation on inner side of ring and along branches. *a*, bone ring; *b*, osteoblasts (enlargement of Figs. 19 and 30 *A*).

A



B

FIG. 21.—*A*. Osteoblasts on both sides of ossifying branches. *a*, branches (enlargement of Fig. 19); *b*, osteoblasts. *B*. Osteoblasts on both sides of ossifying branches (enlargement of Fig. 21 *A*). *a*, osteoblasts; probable origin of osteoblasts from conjoined transformed fibroblasts. *b*, branches.

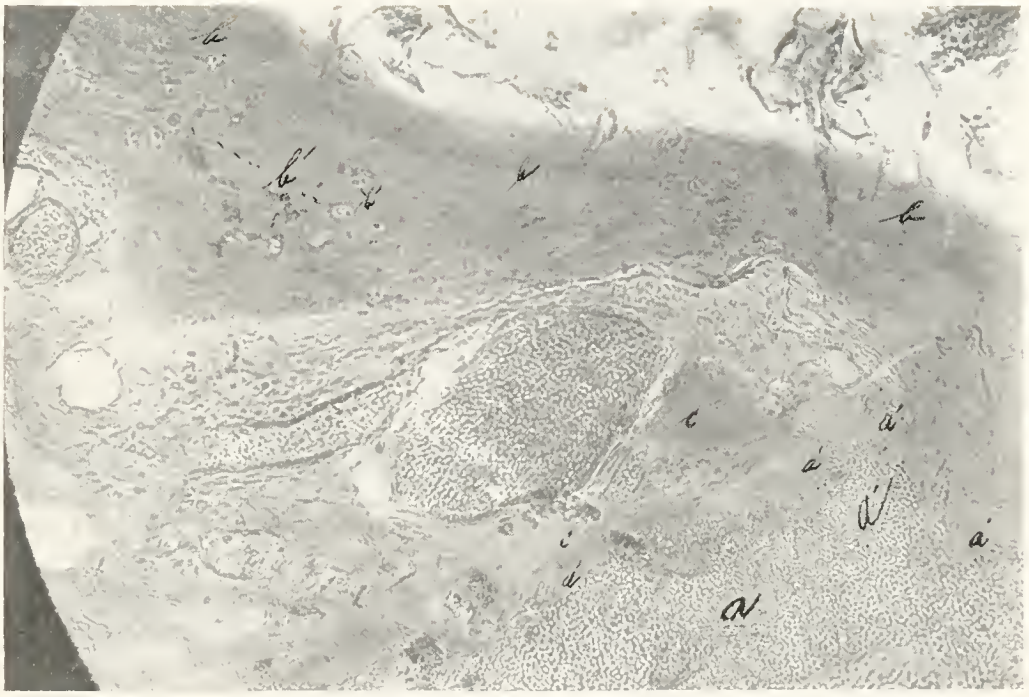


FIG. 22.—Inner part of ring cartilaginous outer osseous—Erythrocytes aiding bone formation. *a*, erythrocytes apparently entering bone formation; *a'*, erythrocytes in close opposition to youngest layer of ring changing shape to spindle form; *b'*, cartilaginous spot; *b*, bone ring.

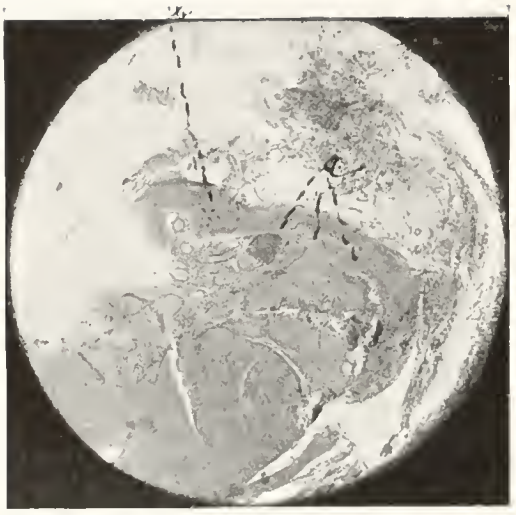


FIG. 23A

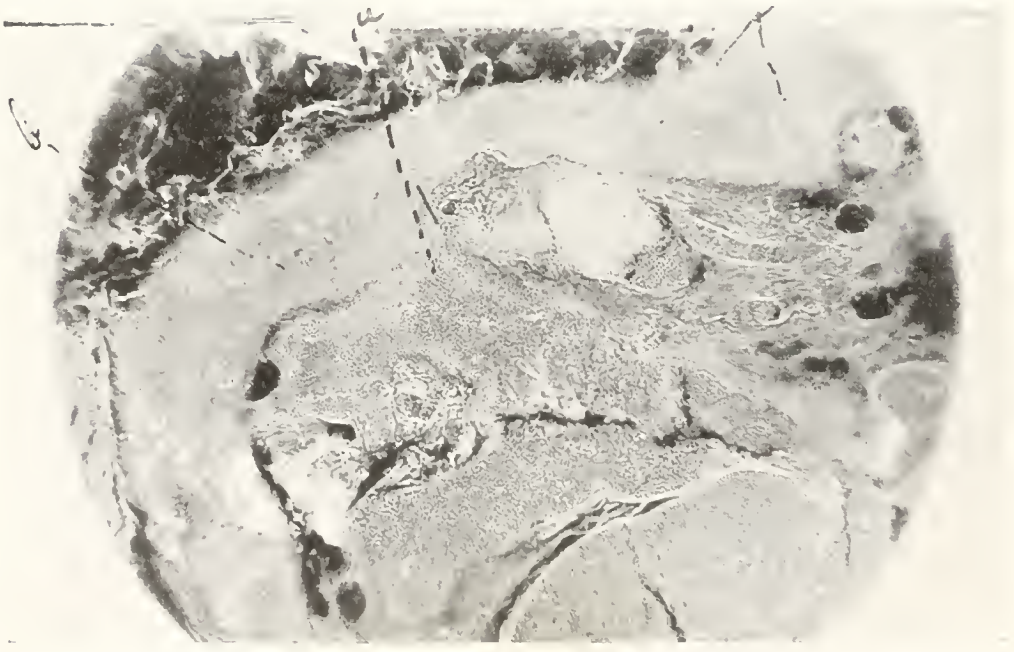


FIG. 23B

C

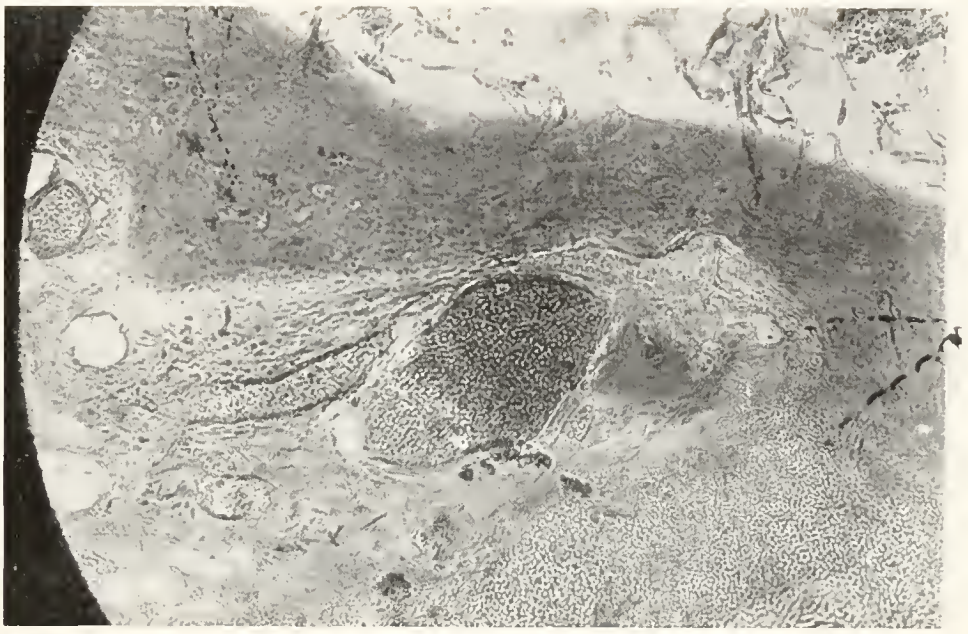
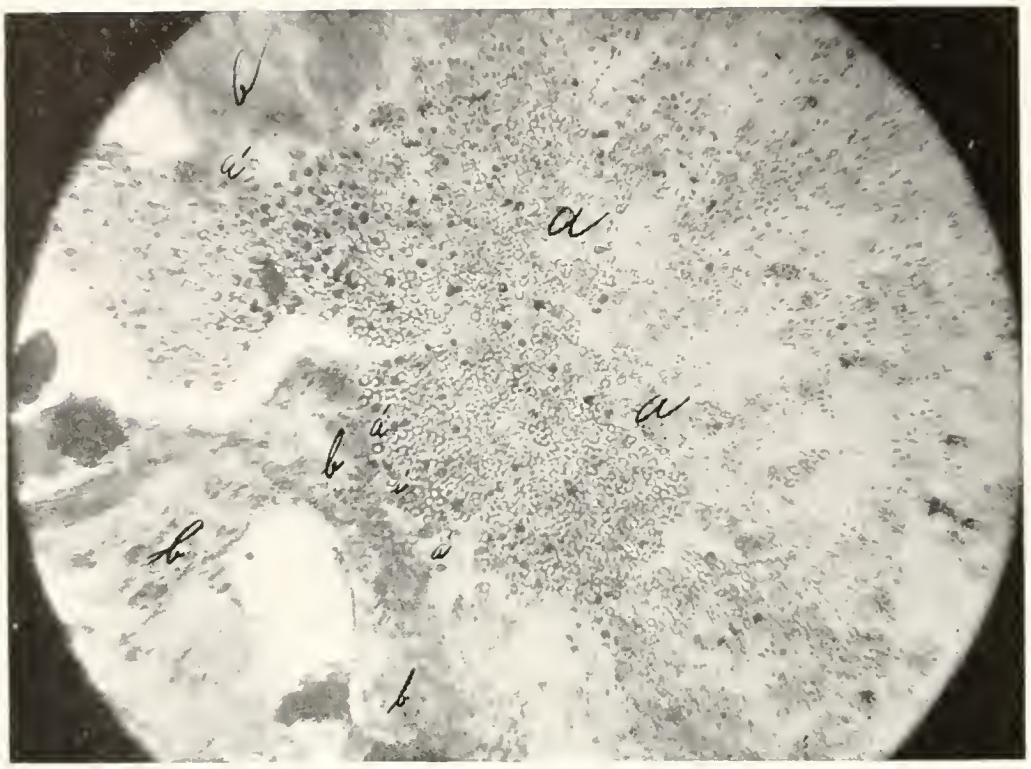


FIG. 23.—Erythrocytes apparently entering into bone formation and remainder of cartilaginous tissue next to osseous formation in ring. *A*, *a*, erythrocytes apparently aiding bone formation; *b*, cartilaginous spot (oc. II, obj. 16). *B*, *a*, erythrocytes apparently entering bone formation; *b*, cartilaginous spot (oc. VI, obj. 8). (Enlargement of *A*.) *C*, *a*, erythrocytes apparently changing into fibroblasts; *b*, cartilaginous spot (oc. VI, obj. 4).

FIG. 24.—Apparent erythrocyte participation in bone formation of branch and ring. *a*, erythrocytes; *a'*, change of form of erythrocytes to spindle shape as they get into contact with the youngest osteogenetic layer of the ring and branches; *b*, bone ring; *c*, branch (very early stage).

B



C

FIG. 26.—*B*. Apparent erythrocyte participation in building of branches in very early stage of formation (enlargement of Fig 26 *C*). *a*, erythrocytes; *a'*, same entering into building of branches; *b*, branches in very early stage of formation. *C*. Apparent erythrocyte participation in building of ring and branches in early stages of formation somewhat later than Fig. 26 *B*. *a*, erythrocytes; *a'*, erythrocytes in apparent participation of building of ring and branches in early stage of formation; *b*, ring.

a spur about 3 cm. long at its upper part. He was under daily observation until February 15, 1895, when the surgeons discovered an osteoma in the right adductor medius. It had formed unbeknown to the patient, occupying the same position as on the left and evidently from the same cause.

Duems[34] expresses his belief, founded on the observation of his cases, that the irritation of the connective tissue from the hemorrhagic infusion following the trauma is responsible for the ossification.

Bier[35] voices a similar opinion, accusing directly the hemorrhage of being the irritant for bone-formation.

Stempel[36] excised pieces of muscles from a case of myositis ossificans progressiva. His microscopical examination of the early stages always showed a hypertrophy of the fibrillary tissue separating the muscular fibres and an intramuscular hemorrhage, and although he never detected an increased cell infiltration or primary hypertrophy without hemorrhage, this author expresses the opinion that the excessive connective-tissue growth as the primary factor in the process causes the hemorrhage by pressure and later the ossification.

Steinert[37] observed ossification of muscles after hemorrhages into them, with patients whose limbs were paralyzed from cerebral apoplexy. In some of these cases the connection between hemorrhage and ensuing ossification is most striking, in a few others only very probable. In one of the cases a disposition to hemorrhages existed.

In their experimental work Sacerdotti and Frattin[38] found a nearly constant bone-production with bone marrow in kidneys of rabbits after ligation of renal blood-vessels, and consequent hemorrhagic necrosis and ossification throughout the organ. The necrotic tissue is riddled with blood-extravasations, and these give rise to ossification.

Berndt[39] found, in a free ossified muscular tumor extirpated twelve days after a trauma, a hemorrhagic extravasation entirely permeating the muscle and periosteum.

In his two excellent monographs on myositis ossificans progressiva[40] and myositis ossificans traumatica Goto[41] pleads for the recognition of an essential difference in the part taken by the hemorrhagic extravasation of changing into the ossifying tissue of the former disease from that in the latter. Although encountering in all of his early excisions of invaded tissues, either of the progressive or the traumatic, or of the experimentally produced forms of myositis ossificans, the characteristic hemorrhagic extravasation into the affected territory, he considers the hemorrhage of the progressive form a secondary feature induced by the hyperplasia of the connective tissue, which is supposed to finally tear the muscular fibres, together with the small, thin-walled blood-vessels, and thus cause a secondary hemorrhage, an explanation already advanced by Stempel.[42] In the traumatic form, however, Goto

[34] Deutsche militaeraerztl. Ztschr. 1887. No. 16, p. 325.

[35] Beobachtungen ueber Kochenregeneration. Arch. f. kl. Chirurgie. Bd. 100, p. 91.

[36] Op. citat.

[37] Ueber d. sogen. Myos. ossif. & verw. pathol. Verkngsproc. insbes. bei org. Nervkrkhtn. Mithlg. a. d. Grenzgeb. d. Med. & Chir. Bd. 21, 1910, p. 513.

[38] Sulla produzione eteroplasto dell osso Ricerche sperimentali. Giornale della Academ. di Med. di Toreno. Vol. vii, 1901; fasc. 12 & Virch. Arch. 1902, Bd. 168.

[39] Arch. f. kl. Chirurgie. Bd. 65.

[40] Patholog. & Anatom-klin. Studien ueb. d. sogen. Myositis ossif. progr. Arch. f. klin. Chir. Bd. C, p. 730.

[41] Die sogen. Myositis. ossif. circumscripta traumatica. Eine chirurg. experimentelle Studie Mitthlgn. aus. d. mediz. Fakultaet d. Kais. Univ. Kyushu, Fukuoka, Japan, Bd. 1, 1914.

[42] Op. citat.

recognizes the primary part of the hemorrhage, and the microscopical examination showing no difference between the two, he considers hemorrhages in the latter the primary cause and essential for bone-growth in consequence of the trauma; and although he constantly found their traces as well in fresh attacks of myositis ossificans progressiva, they are considered by him as a secondary trait only in this disease.

Goto employed for his experimental production of myositis ossificans indirect traumatism of the periosteum by hammer blows upon a portion of the femur, only thinly covered by muscle, and transplantation of periosteum into the muscular substance by either pedicled or free flaps, or of fine pieces chopped in physiological salt solution. They were all successful. He also tried hammer blows, and other traumatisms, with and without intramuscular injections of small quantities of blood, and finally the method of G. Koga[43]—repeated intramuscular injections of 1 per cent. solution of chlorate of lime, 3 cm. to the kgr. weight of rabbit. Of this last series of experiments, solely the ones with chlorate of lime injection showed ossification centres; the ones with the injections of blood alone showed calcification only.

The excisions and examination of these latter took place only thirteen days after the injections, whilst those of the chlorate of lime series were allowed to stay thirty and more days. The question if a longer interval between injection and excision of the blood injections would have given results of ossification must remain unanswered at the present time. The same experiments on guinea pigs showed early X-ray shadows.

Duems-Vulpius[44] approach this quest from an experimental as well as clinical standpoint. Their experiments and their casual observations of these ossifications always showed hemorrhagic extravasations as their precursors.

Among the latest publications on heteroplastic ossifications, two notable papers, Moshkowitz[45] "On the Relation of Angiogenesis to Ossification, etc.," and George W. Outerbridge's[46] "Non-teratomatous Bone-Formation in the Human Ovary," require special attention. Moshkowitz quotes the older Rabl-Schulte doctrine of the specificity of endothelium, which takes it from the series of mesenchymal derivatives and separates it absolutely from the blood should this prove to be of mesodermal origin. Consequently, thinkers who support this view have seen the importance of assigning an entodermal origin to both blood and endothelium. He then endorses the modern theory of angiogenesis (first proposed by Rueckert and Mollier) that blood-vessels form from clefts and spaces within the mesenchyme bounded by the indifferent mesenchyme cells. The clefts and spaces enlarge, fuse together to shape the blood-vessels while the lining mesenchyme cells flatten and form the endothelium. The microscopical specimens of Moshkowitz lend great force to this theory. He ascribes to blood-vessels the important part in the process of ossification in bringing a dead or inert tissue into heteroplastic activity; and in describing further how fibroblasts later become endothelium he shows that normal and pathological ossification are based upon identical features.

[43] Koga—Kyota Igaku Zassi. Bd. IX, 1912, h. 2 and 3.

[44] Op. citat.

[45] The Relation of Angiogenesis to Ossification. Based upon the study of five cases of Calcification and Ossification of the Ovary. Johns Hopkins Hospital Bulletin, No. 301, March, 1916.

[46] *American Journal of the Medic. Sciences,* No. 531, p. 868; June, 1916.

In his conclusions, he reiterates "the importance of circumscribed lime deposits for the formation of primary Haversian canals, accomplished by the genesis of an active mesoblastic tissue, both around the surface and within the interior of such deposits. This mesoblastic tissue is derived from the adjacent blood-vessels and the predominant activity in the development of new blood-vessels." . . . .

The development of new blood-vessels affords the keynote to the interpretation in terms of cellular ontogeny of the process of ossification. The histological constituents which enter into the formation of new blood-vessels are the progenitors of all the histological compounds of osseous tissue. In other words, blood-vessels, osteoblasts, bone cells and marrow (in large part at least) are merely differentiations of the mesenchymal cell-unit. After having demonstrated that his specimens furnish strong corroboration of the adaptive or mesenchymal theory of angiogenesis, and to the theory of the non-specificity of endothelium, he adds that ossification does not occur without preliminary calcification, and calcification occurs only in dead tissues. We quoted Moshkowitz more extensively, as our own specimens show many resemblances to his, after calcification has occurred. Outerbridge gives virtually identical pictures but does not enter into their interpretations, but simply chronicles the calcification and final ossification.

Although Haga and Fujimura[47] do not ascribe a basic importance to the primary hemorrhage in the traumatic myositis ossificans, the reports of their microscopical findings in extirpated specimens of their cases variously mention in every one the presence of blood-extravasations; as "capillaries in the intermuscular connective tissue are torn and extravasation of blood has taken place . . . numerous and large hemorrhagic extravasation and mighty development of new capillary vessels . . . goodly sized blood extravasation in the intramuscular connective tissue . . . here and there blood extravasates and neoformation of capillaries . . :" and thus permit a different view of their importance from that of the authors.

Van Arsdale[48] published a very interesting and important case of an ossifying hæmatoma, important for this question of participation of free blood in the act of ossification. A young man, 22 years old, received about the middle of April, 1891, severe blows on his right arm. The region immediately below the insertion of the deltoid was the seat of a large, soft fluctuating tumor, showing an increase of circumference over its fellow of 10 cm. The skin over the swelling was bluish and the parts tender on pressure. When seen again, May 20, the circumference of the right arm was diminished 6 cms., tumor of bony hardness throughout and apices harder than middle. Presented on May 27 to Surgical Society of New York; diagnosis, ossifying hæmatoma. Operation June 22. External tumor 9 cm. and 3 cm. broad or high, external coat or shell nearly 1½ cm. thick; contains cavity filled with partly coagulated blood. Bony hardness of the wall most marked over periphery of tumor. Induration then gradually decreased toward central cavity.

The same course, from calcification to ossification, as seen in humans, was

[47] Arch. f. kl. Chir. Bd. 72, Hft. 1.

[48] Annals of Surgery, 1893.

observed in the one case published as such from animal pathology. Lorge[49] dissected an anatomy horse which showed the symptoms of progressive myositis ossificans. The horse showed in the connective tissue surrounding the smaller muscular bundles (perimysium internum) a great number of fine needles resembling cement, which made a noise when the knife passed over them. The affected muscular fibres had a pale yellowish color; they were atrophic and their contents partly fattily degenerated.

The diseased muscles were distributed all over from head to feet. In the tongue the mass could be kneaded with the fingers; in other places it was so firm that it could be crushed only with difficulty. Microscopical examination showed that the normal striation of the diseased muscles had disappeared and in their place were granular or dark dots composed of bone cells. The external cellular sheath of the muscle did not show anything of a pathological calcification. According to the views of Lorge, "the calcareous deposit in the inner sheath of the muscle was the first, the atrophy and fatty degeneration of the muscle fibre the second stage of a disease, which is rarely observed with humans and has been named progressive ossifying myositis by Professor Dusch, of Heidelberg."

Another animal case not yet published is one observed by Prof. Dr. K. F. Meyer, of the Hooper Foundation for Medical Research of Medical School, University of California. While showing my slides to the Doctor he was reminded of the case of a dog examined by him in Philadelphia in 1912 as Director of the Pathological Laboratory, Pennsylvania State Live Stock Sanitary Board, University of Pennsylvania. From comparing our notes and pathological findings we both came to the conclusion that this dog, the exact diagnosis of whose disease had been rather baffling, had been suffering from myositis ossificans progressiva, and Doctor Meyer was kind enough to permit me to incorporate his hitherto unpublished manuscript in my treatise. I give it largely in its original form, with a few changes made by me relative to its new viewpoint and submitted to and approved by Doctor Meyer.

In November, 1912, through the courtesy of Doctors Cunningham, Hoskins and Jarrett, of the Pennsylvania State Live Stock Sanitary Board, a dead setter dog was sent into the Pathological Laboratory which showed on all four legs a most remarkable anomaly which had developed gradually. Unfortunately, the dog had died the day previous, and therefore the carcass when brought to autopsy was in a fairly advanced stage of decomposition. Before giving the autopsy findings, I will briefly relate the clinical history given to me by the owner of the dog (Doctor Cunningham):

Setter dog; male; was born in May, 1909, and was one of a litter of fourteen. Some of the pups were killed accidentally by the mother. Seven were raised. They were all extremely nervous. At the age of four to five months the pup in question was lame in the left hind leg. It was found that the left hock region was swollen and on palpation very tender. The animal was treated with Fowler's solution for about one month, at the end of which time the swelling disappeared and the dog developed normally to a typical English setter and at the age of one year won two first prizes. At the age of eighteen months the dog became listless, would lie in the corner and refused to play, which was strange, as the dog had always shown unusual vivacity. At the age of nineteen months the swelling on the hock again appeared, was tender on palpation and soon extended above and below the hock. The dog became extremely lame and was most of the time dragging his legs under him. It was again treated with Fowler's solution, and about two months later a similar swelling started in the right hock, and at the age of twenty-two months both hind legs were enormously enlarged. A careful examination revealed that the joints were absolutely

[49] *Annales de Médecine vétérinaire;* publ. à Bruxelles XXme année Mars.-Mai, 1871, ref. Repertorium d. Thirhlkde, Hering.

normal. The general condition of health was perfect; the appetite was good; no fever was ever noticed.

In the spring of 1912 a similar swelling started gradually, beginning on the metacarpus and elbow on the front legs. The dog started a very troublesome cough. This condition remained the same until November, 1912. He was able to walk by dragging his legs after him, but he had always to be assisted when walking up or down stairs. Two days before his death the cough was very bad and temperatures from 103 to 106 were noted. He died with the symptoms of heart paralysis November 19, 1912.

The autopsy was performed by me on November 20, 1912, and the following is a condensed report of the essential findings only:

Setter dog; male; 2½ years old. Emaciation; all four legs hard, tremendously enlarged and heavy. Fat tissue has nearly disappeared. Muscles on extremities atrophic, pale and ossified. Along the extremities the skin can only be removed with difficulty on account of a heavy layer of dense, hard, whitish connective tissue. Along the shoulder-blade at the insertion of the muscles, hard, nodular, fibrous, callous-like connective-tissue layers.

In both pleural cavities there is a small amount of reddish fluid. Costal pleura slightly reddish.

The right apical lobe is attached to the costal pleura by several broad connective-tissue bridges. All lobes of the lungs are œdematous and soft. The right upper lobe shows a tumor size 15½x19½ cm. distinctly protruding above the surface of the healthy lung tissue. Its color is whitish and in certain places grayish.

The consistency of this tumor is dense and hard. On section there is a connective-tissue capsule of about 6 mm., a whitish, yellowish tissue meshwork, which contains many cheesy, pus-like areas. The bronchi leading to this portion contain granular pus and a few superficial ulcerations. All the other lobes of the lungs contain in the bronchi a serous reddish fluid. The tissue is soft and friable.

The left and right bronchial lymph-nodes are enlarged and contain numerous yellowish nodules, which are in places also cheesy. In the trachea and bronchi a small amount of granular mucus intermixed with pus flakes. The vocal cords and the epiglottis are somewhat thickened and hard.

An examination of the extremities revealed the following conditions: The left and right thoracic limbs show the length of the ulna about 22 cm., a circumference of 22½ cm. and a thickness of 6 cm. The elbow and carpal joint are immobile. The metacarpal and interphalangeal joints are, under restriction, mobile. On incision the enlarged parts of the ulna show a fibrous, nodular, gritty, bone-like material. Both pelvic limbs are extremely enlarged, but remarkably flattened. The following figures will indicate the conditions:

The length of the right tibia is 25 cm. From the middle of tarsus to the end of the third toe, 18 cm. The diameter of the oval-shaped tibial circumference is 8 cm., and 6 cm. thick, and is considerably flattened in the middle. The circumference is 21.5 cm. The circumference of the metatarsal bones is 25 cm., and the diameter of the region of the tarsal bones is 10 cm. The second toe has a length of 3 cm. and a circumference of 5 cm., and is extremely plump. The structure of the subcutis is the same as that described for the fore-limbs.

No swellings or enlargements of any kind were noticed on the head, around the spinal column or the tail.

The left thoracic and the right pelvic limbs, together with the entire skeleton, were carefully macerated. The right thoracic and left pelvic limbs were fixed in Mueller's solution. The tumor in the apical lobe was fixed in formalin.

The maceration of the skeleton revealed the following pathological changes:

The supraoccipital and the condyloid fossa show slight roughening. The condyles

of the mandible show small, wart-like roughenings. All the bones of the head are somewhat porous.

The left shoulder-blade is light when compared with those of normal dogs; somewhat porous, but normal in shape. The posterior angle shows plump, thick, hard swelling, size 2½ x 4 cm. (See plate 1, 2, 3.) The edges are nodular, wart-like. The acromion is enlarged, flattened and porous. The scapular notch shows small, wart-like, spongy exostoses. The scapulo-humeral joint is absolutely smooth and normal. The humerus is normal in length, but extremely plump; shows on the crest a flat, leaf-like growth of bone tissue, which extends over the shaft, leaving a space free for the atrophic biceps muscle. Both the external and the internal condyloid crest, the external and internal condyle, the olecranon fossa, are covered by heavy (3 cm.), thick, spongy, wart-like osteophytes, resembling on the internal condyle the picture of a sponge. Both articular ends of the humerus are normal. The bone tissue is extremely spongy and soft. Above the condyles is a small hole connecting the coronoid fossa with the olecranon fossa and is caused by osteophytes on the proc. anconeus of the ulna.

Both ulna and tibia are covered with most luxuriant osteophytes. Their length appears about normal as compared with dogs of the same age. The existence of the ulna can only be determined on the proximal joint ends, where the radius is still separated from the ulna by a small groove. Otherwise the entire ulna is buried in the mass of new bone tissue. The thickness of this neoformation examined from the anterior side is about 5½ cm.; spongy, irregular masses of bone tissue surround the entire radius, leaving only on the internal side a round groove for the localization of the deep flexor and the flexor carpi internus. The joint ends are entirely invisible when mounted, but show distinct pseudo-arthrosis of the osteophytes. A similar pseudo-arthrosis is seen at the proximal end of the osteophytic growth and the osteophyte above the coronoid fossa.

The carpal bones are invisible on account of the masses of osteophytes. On the posterior view they can still with some difficulty be distinguished, as there the osteophytes have left a flat, oval channel for the passage of the flexor tendons. The metacarpal bones are uniformly covered in the anterior portion by an arborescent wart-like growth of osteophytes. The third and fourth bones are separated by a cleft. From the fifth and the first metacarpal bones the osteophytes grow over the posterior part of the metacarpus, forming the roof of the channel mentioned above. The osteophytes on the metacarpal bones show pseudo-arthrosis with the osteophytes of the carpal bones, the ulna and the radius. We have two masses of osteophytes, therefore, those of the radius and ulna and those of the carpal and metacarpal bones, which are connected by joints.

The first phalanges are plump; those of the fifth, second and first digits show the osteophytes on the external side. The second phalanx of all digits is chunky and shows wart-like osteophytes growing on the back of the phalanx. The wing and coronary ridge on the third phalanx are also plump and show crest-like osteophytes surrounding the entire bone. Only the ungual surface is entirely smooth, somewhat porous. All of the joint surfaces are smooth and unaffected. The volar sesamoids were lost during the process of maceration.

The pelvis appears normal, is somewhat porous; around the psoas tubercle and the iliopectineal line there are several wart-like osteophytes.

The femur is of normal length; shows similar conditions to the humerus; the proximal and distal joint-ends are normal.

The patella and the sesamoid bones are covered with osteophytes which are connected by pseudo-articulations.

The tibia and fibula are remarkably enlarged and show most of the lesions that were observed in the thoracic limb. Here also a space is left open for the flexor tendons, the lesions on the tarsus and metatarsus and digits are nearly identical with those just described for the thoracic limb, with the exception that there is a marked

lateral compression, probably due to the fact that the dog was lying constantly on the hind legs.

Most of the lesions are therefore seen on both extremities. They are characterized by extremely massive apposition of bone which is located around the compact masses of the bones, selecting frequently the places of the insertion of the tendons.

A chemical analysis of some of the osteophytes on the radius, and, for comparison, from the shoulder-blade, was made. I am indebted for the analysis to Dr. John Marshall, of the University of Pennsylvania.

CHEMICAL COMPOSITION OF DRIED BONE

	Normal	Exostosis	Shoulder-blade
Calcium phosphate, $Ca_3(PO_4)_2$	57.35%	53.98%	52.71%
Magnesium phosphate $Mg_3(PO_4)_2$	2.05%	2.75%	2.18%
Carbonate, fluoride, and chloride of calcium	3.45%	3.04%	3.32%
Organic matter (ostein)	33.30%	31.91%	32.58%
Water at 105°–120° C	.....	8.32%	9.21%

By the above table it is shown that the minerals are uniformly reduced as well as the ostein. A slight increase of water is noted and conditions are prevailing here which resemble somewhat those found in osteomalacia.

The microscopical examination of sections of the lung tumor, stained with hæmatoxylin, eosin, Van Gieson, etc., show:

A thick layer of connective tissue containing a few capillaries. Below this layer is seen the tumor tissue of alveolar or papillomatous epithelium. Connective tissue septa of varying thicknesses form a net-like stratum; the free spaces, tubules or alveolæ are coated by single or stratified layers of fairly regular epithelial cells and contain débris, desquamated cells and coagulated masses.

The size of the alveolæ is varying; they are mostly tubule-like; in some places they are glandular and resemble adenomatous or papillomatous structures. The epithelial cells are squamous, with large vesicular nuclei; sometimes they are narrow and cylindrical; in most alveolæ they are in single layers. Most of the cells have smooth contours; some, however, show filament-like protoplasmatic projections. The type is apparently one of metaplastic pulmonary epithelium. It is impossible to determine whether alveolar epithelium of the lung, or whether metaplastic columnar epithelium of the bronchial lining is the substratum from which these tumor cells have developed. In specially stained sections no mucin-like material could be seen, so that I feel justified in excluding the possibility of the tumor cells having originated from the mucous bronchial glands.

In the depth of the tumor the tissues are to a large degree necrotic and consist of material which stains deeply with eosin; some areas of liquefaction are distinctly seen as indicated by the presence of numerous leucocytes and lymphocytes.

The lymph-nodes showed the same papillomatous arrangement of the same metaplastic epithelial cells. A careful search for tubercular lesions was negative. The lung tumor can therefore be diagnosed as a primary lung carcinoma with secondary metamorphosis and metastases in the bronchial lymph-nodes. It was impossible to determine if the tumor was of *alveolar* or of *bronchial origin*.

Sections of the bone proliferations which were fixed in Mueller's solution and decalcified with nitric acid and embedded in celloidin show on the left fibula (for example) that the entire bone material directly under the derma is covered by fibrous connective-tissue layers. The osteophytes in form of lamellæ and trabecula are rich in blood-vessels and Haversian canals. Some of the walls of the blood-vessels are abnormally thick. Fibrils of connective tissues cross the muscles in different directions and compress the fibres. Between the osteophytes is a soft, succulent connective-tissue mass.

The compact substance of the fibula is extremely reduced in thickness—and it is difficult to realize what is originally bone and what is osteophyte. The Haversian canals are very remarkably enlarged; one notices groups of osteoclasts and giant cells scattered irregularly. The process here is of the usual character of bone resorption.

Below this zone similar trabecules grow into the marrow. The structure and shape are similar to the periosteal osteophytes. Dense connective-tissue fibres show layers of osteoid tissue which is surrounded by spindle-shaped cells resembling osteoblasts. The process of active bone apposition is quite apparent. Lack of comparative material prevented a further study of this enchondral ossification.

In view of a recent revision of the data of this case I do not doubt it to be one of myositis ossificans progressiva in the dog. The clinical history is an exact counterpart of many of the typical histories of human cases in the foregoing compilation (*vide* cases 15, 17, 23, 25 and many others), the more extended ossification having been arrested by an early death due to complication with the carcinoma in the lung. The pathological changes in the osseous system are significant of the disease. The ossification along the muscular tracks and insertions involving and supplanting these structures, the comparative freedom of the articulations, together with the spongious character of the new bone formation, are all features of this disease. A very characteristic symptom marking this case undeniably as one of myositis ossificans progressiva is the microdactylic state of the thumb of the photographed thoracic extremity. The end phalanx was lost in the process of maceration, but the first metacarpal is less than half of the normal size and the first phalanx also is considerably shortened. At the time of the autopsy the diagnosis of myositis ossificans progressiva was not taken into consideration and therefore no attention was paid to the fingers and toes of the other extremities; but the photograph of the macerated foot shows the condition distinctly so as to remove all doubt, at least as to this extremity. The very rare similar observations in the literature of animal pathology appear from this viewpoint in a new light.

In 1890, Kitt[50] observed a case which, according to his description, resembles my specimens in many respects. The exception was the exceedingly prominent lesions on the mandible very frequent in human cases of myositis ossificans progressiva. Perhaps inasmuch as the dog had been destroyed in perfect health, the lesions on the limbs could not develop to the same degree as those seen in the case under discussion. The dog was one year old. Kitt[51] explained the condition as an abnormal diathesis of bone formation, and inasmuch as the ossification started at the insertion of the muscles, he diagnosed his case as a multiple myopathic hyperplasia of the bones.

Liénaux[52] saw two old dogs with distinct enlargement of the thoracic and pelvic limbs. He could autopsy, however, one animal only. The pictures and descriptions correspond with my observations only that the extent of the process is a less marked one than in my case. In another case, of which he found only the skeleton in the

[50] Kitt: Lehrbuch der pathologischen Anatomie der Haustiere, I, 1910, 4th edit., p. 388.

[51] Symmetrische multiple Hyperostosis & Exostosis bei einem Hunde tahresbericht der kgl. tierarztlichen Hochschule, 1890.

[52] Liénaux: Apropos de l'ostéitisme, ostéite déformante du chien. Ann. de médecine vétérinaire. Mars, 1899.

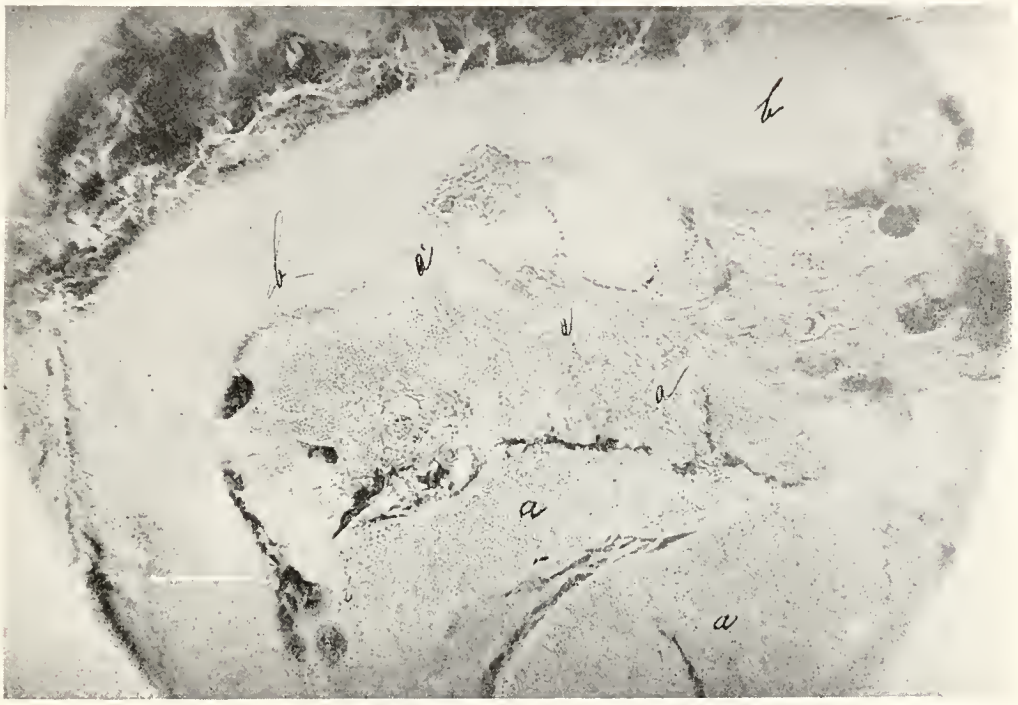


FIG. 27.—Apparent erythrocyte participation in bone formation and differentiation of osseous and cartilaginous parts of ring. *a*, Erythrocytes; *b*, cartilaginous part of ring; *c*, osseous part.

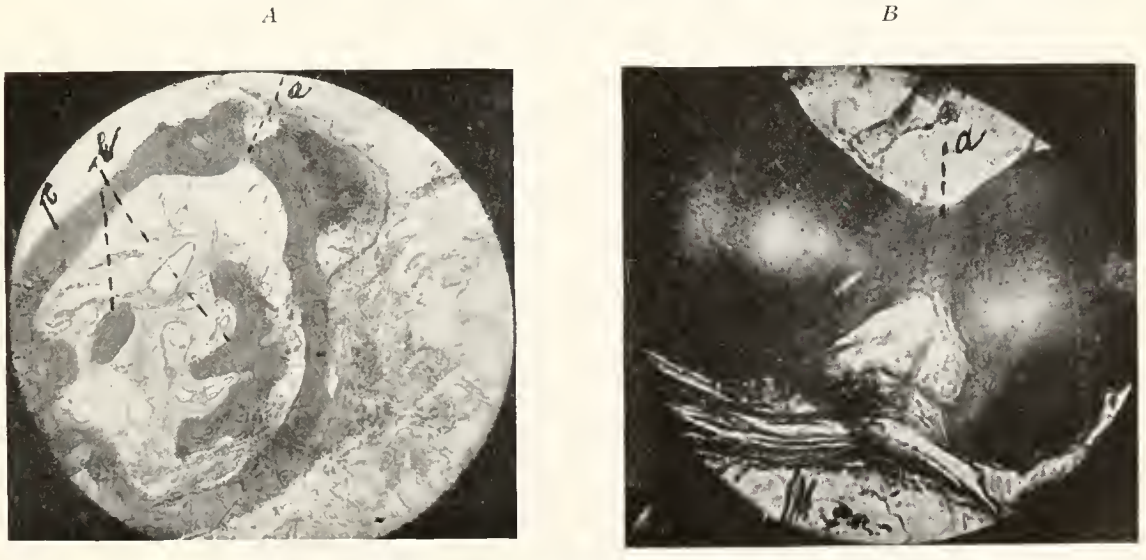


FIG. 28.—Interior osseous branches and islets forming and a connective-tissue bridge between bony ends of ring. *A*, *a*, connective-tissue bridge; *b*, branches and islets; *c*, outer ring (oc. 6 obj. 16). *B*, *a*, connective-tissue bridge of *A* (oc. 6, obj. 4).

FIG. 29.—Bone and bone-marrow. *A*, *a*, outer ring. *b*, bone marrow (oc. 2, obj. a'7). *B*, *a*, outer ring; *b*, bone-marrow (oc. 2, obj. 16). *C*, *a*, branches; *b*; fibroblasts and capillaries (oc. 2, obj. 4). (Enlargement of inner part of *A*.)

FIG. 30.—Cartilaginous development of bone. Cartilaginous tissue at inner border of osseous ring. *A*. *a*, ossified outer part; *b*, cartilaginous part; *c*, epidermis (oc. 2, obj. 16). *B*. *a*, ossified part; *b*, cartilaginous part (oc. 2, obj. 8). (Enlargement of *A*.)

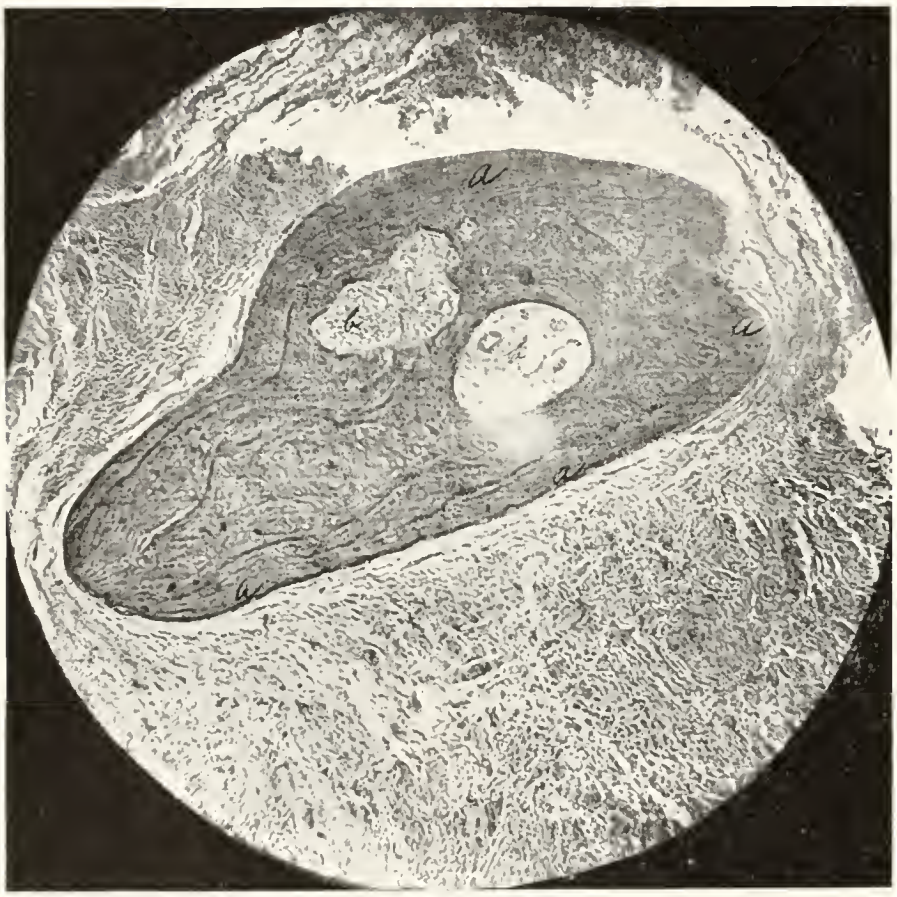


FIG. 34.—*D*. Bone in late stage of solidification with small remainders of hemorrhagic enclosure. *a*, outer bone layers; *b*, remainder of hemorrhagic enclosure. (Enlargement of Fig. 34 *C*.)

museum at Brussels, were seen similar symmetrical exostoses. These cases were diagnosed by him as ostitis deformans. Ten years later, on account of some further observation along the same subject and influenced by the work of Alamartine[53], the same writer revised his early conception of the bone lesion and considered it to be identical with the so-called "osteo-arthopathia hypertrophiante" in man. Ball and Alamartine[54] had observed a tubercular dog which had the same macroscopic and microscopic processes as those seen by Liénaux, and therefore it suggested itself to Liénaux[55] that the term "osteoarthropathy" would be better than "diffuse osteo-periostitis" or "ostitis deformans."

The few cases described by Kitt and Liénaux show great analogies with my case. The processes, however, are not as far advanced in either case, and there are also differences in the ages of the dogs affected. The animal seen by Kitt was one year old; those reported on by Liénaux were all over ten years old when first seen. Aside from these minor differences, which also occur frequently in human patients, the cases are identical with mine so far as concerns the anatomical lesions of the bone system.

In none of these cases a satisfactory clinical or causal explanation has been given; the tuberculosis in Ball and Alamartine's case was an accidental complication, and they all belong most probably to and should be classified as myositis ossificans progressiva of the dog.

In order to arrive at a better understanding of the basic features of our malady, it appears advisable to me, before we enter into a discussion of the peculiar and specific characteristics of the progressive ossifying myositis, that we should now consider collectively all those traits it possesses in common with its near relative, the traumatic myositis ossificans and its experimental imitation. They all center around the pathologic anatomical structural pictures and bone-forming process; they are identical in appearance and modes of procedure in both affections.

In fixing its place among the pathological entities it occurs at once that Virchow's view of its being a multiple exostosis must be abandoned in spite of the master's word, and the free growth conceded within the confines of the muscle and its surrounding connective tissue.

We have the research work of many authors, including our own case, to establish the evidence of independent growth of bone tissue within the muscle bundles and their connective-tissue envelopings and dividing partitions.

That question also as to the primary point of attack, still indicated by the name myositis, may be considered settled. Every observer who has excised a piece of the affected tissue at an early stage of the local pathological change has found the inter- and intramuscular connective tissue to be the primary

[53] Alamartine: Ostéoarthropathies hypertrophiantes d'origine tuberculeuse. Revue de chirurgie 35, 1907, p. 992.

[54] Ball & Alamartine: Pathologie et Ostéoarthropathie hypertrophiante d'origine tuberculeuse chez l'homme et chez le chien. Revue de chirurgie, 1908.

[55] Liénaux: Remarque au sujet de la nature de l'ostéo-arthropathie hypertrophiante. Bull. de l'Académie Royale de Médecine de Belgique. 27 février, 1909.

seat of attack, even before my discovery of isolated bone-tissue in the skin removed the possibility of doubt. In both diseases the muscle fibres are the secondary sufferers and succumb only to the increasing pressure and embodying force of the growing ossification. While these two questions are easily absolved, we encounter a greater difficulty in order to arrive at a satisfactory result in establishing the manner and the identity of the bone-forming process.

Although the oldest attempt to explain the heterotopic ossification by a metabolic retention of lime has been refuted by the exact modern investigations of both Burgerhout[56] and Austin[57], the fact that its presence as a local deposit is an essential condition for bone-formation continues to be valid. The truth of this assertion has been proven by the experimental work of Dibbelt[58] through calcium supply by way of the alimentary canal, and by hypodermic injection of 1 per cent. solution of chloride of calcium of Koga[59], this latter successfully repeated by Goto[60]. Many other authors confirm it by their observations on humans and point at the initial hemorrhage as the source of the calcium supply in spite of its rather low percentage in the blood. The idea of looking to the blood as the first cause of ossification, either as a direct irritant for its initiative stages or as a chemical one through its lime deposit, lies very close if one considers that in cases of traumatic ossifying myositis the first effect of the trauma is always a hemorrhage into the injured tissues.

We may dispense in these cases with a serious consideration of the periosteal participation, as only the entirely severed and intramuscularly implanted periosteum gives rise to a free bone-formation, whilst the flap always shows the new-formed bone in connection with the underlying skeletal bone, and it would be more than unreasonable to presume that the great majority of traumatisms responsible for a case of skeletal-free myositis ossificans tear off a piece of periosteum and transplant it into the adjoining muscular tissue.

Many clinicians, as we have seen, were able to witness the growth of the newly-formed bone directly from the initial hemorrhage to the final extirpation of the finished product. The well-known Schede blood-clot for the filling of bone cavities has often served its purpose and the microscopical pictures of early stages of traumatic as well as progressive cases show so constantly the more or less well-preserved remnants of an infiltrating blood-extravasation close to or surrounding the various stages of ossification, from earliest connective-tissue hypertrophy and calcification to complete ossification, that the causative connection becomes irresistibly impressive. Such an impression, gradually rising to a firm conviction, I have also

[56] Op. cit.
[57] Op. cit.
[58] Op. cit.
[59] Op. cit.
[60] Op. cit.

gained from the study of my slides, and in their description and illustration I have tried to convey this conviction to my readers. The various anatomical regions show the progress of bone-formation on different paths; osteoblasts with a fibrous or cellular cartilaginous route in the intra- and intermuscular connective tissue to direct calcification and osseous growth in the hypertrophic fibrous connective tissue of the skin, all starting from the primary blood clot. But with even the fullest admission of the blood's causative ossifying action, we have moved but one step farther toward the solution of the mysterious problem. The question arises now, what are the determining conditions to induce in some cases an ossification after a subcutaneous tissue-invading hemorrhage, and in the vast majority of other similar traumatic cases simply an uninteresting and uninterrupted absorption?

In the description of my microscopical pictures I drew attention to a tissue zone surrounding a developing ossifying part, which zone refused to take the stain well, and with its peculiarly indistinct cell and tissue differentiation showed the unmistakable picture of tissue-necrosis.

A similar area, although not specially mentioned as such, may be seen in Goto's microphotographs of his experimental myositis ossificans produced by means of the chloride of calcium injections.

This tissue-necrosis, following the initial hemorrhage, we believe to be another determining factor for the comparatively extremely rare sequel of ossification to muscular traumatism. This tissue-necrosis, as a preliminary condition to calcification and later ossification, is mentioned by Rohmer[61] in his description of bone-formation in focal endocarditic and endarteritic calcareous necrotic ulceration, of which latter Moenckeberg[62] met with ten in 100 cases of senile arteriosclerosis. Pollak[63] found that the ossification took place in all his cases around calcified necrotic foci in the lungs; and let us recall here that local calcifications of necrotic caseous foci are not infrequently seen in old ulcerative pulmonary processes.

Sacerdotti and Frattin[64] produced ossification in secondary necrotic foci of the kidneys, the blood-vessels of which they had ligated, and thereby caused primary hemorrhages. Liek[65] obtained identical results.

J. F. Poscharissky[66] examined heteroplastic osteogenesis in lungs, heart, blood-vessels, lymphatic glands, brain, eye, uterus, liver, thyroid gland, and always met with regressive metamorphosis (necrotic foci) as the primary incentive for calcification which was followed by ossification. He found that there exists a kind of an antagonism between the necrotic centre and the living periphery of the affected tissue nodule, which is being demonstrated by an effort of the connective tissue to enter into the necrotic centre,

[61] Op. citat.

[62] Ueber Knochenbildgn. in d. Arterienwand. Virch. Arch., Bd. 167, 1902.

[63] Op. citat.

[64] Op. citat.

[65] Experimenteller Beitrag. z. Frage d. heterogenen. Knochenbld. Arch. f. kl. Chirurg., Bd. 80, p. 279.

[66] Beitraege (Ziegler's) z. pathol. Anat. & allgem. Pathologie, Bd. xxxviii, p. 135.

shows a constant tendency to grow. He concludes that osteogenesis may occur in any organ of the human body if there exists a necrosis of the tissue, or at least a sclerosis so intense that it is interfering with the natural blood supply and accompanied by an incrustation with calcium salts. In all his observations Poscharissky never encountered the formation of cartilaginous tissue.

A. Barth shows, in his experiments [67] on bone implantation, new growth of bone only took place after a soil of dead tissue had been provided for it and that[68] calcium favors rapid ossification.

S. Ehrlich[69] holds that the iron when available (from hemorrhages) serves as a mordant for the calcium which is subsequently deposited.

H. Gideon Wells[70] finds as a result of his own and others' experiments that calcification, as the preliminary stage of pathologic ossification, takes place only in dead (necrotic) tissue or in such of low vitality and insufficient circulation. Physiologically, calcification in old age takes place also in ill-nourished tissue, like the laryngeal or costal cartilages.

He gives the amount of calcium contained in blood (serum) of mammals as from 0.11 to 0.131 per cent.,[71] about two to four times as much as is soluble in water.

It may be seen that his conclusion is well supported by pathologic-anatomical evidence. The decision which of the two, the hemorrhage as calcium supplier, or the necrosis as the stimulating and receptive soil for calcification, is the more important for the final ossification, seems impossible to decide at present.

Goto's[72] experiments on rabbits and guinea pigs, in producing intramuscular hemorrhages by severe hammering of the quadriceps femoris never resulted in ossification; the very early excision after the traumatism showed calcifications of the torn muscular fibres amid the fresh hemorrhages; later excisions showed complete resorption of the hemorrhage and the destroyed tissue, in their stead simply a replacing connective-tissue hypertrophy. By the daily injections of 3 cm. of 1 per cent. chloride of lime solution into the quadriceps, continued for thirty consecutive days, bone-formation could be produced. A shorter period, however (eight days), only showed conditions similar to the early traumatic pictures. An unbiased interpretation of these experiments still leaves open the question of the more important factor, lime or necrosis. The lime supply alone by blood extravasation does not seem sufficient to induce the entire range leading to ossification if a reparative

[67] Beitraege (Ziegler's) z. pathol. Anatomie & allgem. Pathol. 1895, Bd. xvii, p. 65.

[68] Ueber kstl. Erzeugg. v. Knochgew. & ueber d. Ziele d. Osteoplastik. Berl. kl. W., 896, p. 8.

[69] Eisen & Kalkimpregnation in menschlichen Geweben. Centralbl. f. allg. Pathologie, 1906, xvii. p. 177.

[70] Calcification and Ossification. Archives of Internal Medicine, vol. vii, June, 1911, No. 6, p. 721.

[71] Op. cit.

[72] Op. cit.

process intervenes and resorption interrupts the progressive stages; is this reparation, however, prevented by a long continuance of lighter daily traumatisms assisted by the constant addition of calcium supply, both carried on by the intramuscular injections of comparatively large quantities of calcium chloride solution, then bone formation is induced.

The foregoing explanatory attempt, although valid for both the progressive as well as the traumatic ossifying myositis, only speaks for both *after* the occurrence of the initial hemorrhage.

While the cause of this extravasation needs no further explanation in the traumatic form of myositis ossificans, a hemorrhage as forerunner to ossification in the progressive form seems as inexplicable as its constant association with microdactylia as a characteristic symptom.

We have tried to lay the hemorrhage to a deficiency in coagulability of the patient's blood, but the tests we made did not bear us out; coagulation occurred even ahead of the normal time, and the reason for the precursory hemorrhage in myositis progressiva ossificans was not explained by any failure of proper coagulation.

It seems, moreover, apparent that all the strange characteristics of this disease are more or less interdependent and might be treated as due to a common cause. It may be possible to advance our understanding of these connective features by examining the progressive ossifying myositis in comparison with other maladies that offer certain symptoms common to both.

The feature of heredity in this disease has hitherto been treated as negligible, as in all former compilations only one case (No. 70, Faughan and Fanning) of hereditary transmission of disease and deformity had been ascertained. We have found two more instances, one (No. 30) and (No. 81) a case of Gaster, going through three successive generations, grandfather, father and three sons, with a selective preference for the male members of the families, who, besides being victims of the ossifications, showed, every one of them, the marked congenital finger and toe deformities.

This deformity, brachydactylia, occurring independently, has engaged the attention of many writers and aroused great interest on account of certain embryological phylogenetic questions. Brachydactylia may be due to either the shortness of a metacarpal or metatarsal bone, or of one or more phalanges, or of either combined, and it may be appropriate to differentiate them by calling the first brachydactylia, the second brachyphalangia and the third brachydactylphalangia or brachyphalangodactylia.

The two views, as to the causes entering into the production of carpal deformities, are the exogenetic, mechanical, and the endogenetic, embryological. The brilliant results of Tornier[73], in experimentally producing various deformities of hyperdactylia in tritons and other animals possessing similar regenerative force, lent great support to the mechanical explanation

[73] Ueber Hyperdaktylie Regeneration & Vererbung mit Experimenten. Arch. f. Entwicklgsmchk. d. Organismen. Bd. iii, 1896, p. 409.

of such deformities as hyper- and hypodactylia, syndactylia, etc., through the agency of amniotic bands and intra-uterine pressure.

Even for brachydactylia a similar explanation was advanced upon the theory of localized pressure from projections of uterine walls, although a clear conception how such pressure could act in the human as to influence the growth of a single metacarpal bone has never been given. Far less possible appears such a solution for symmetrical brachydactylia of one or more metacarpal bones, and we shall try and briefly give our reasons for rejecting this theory for brachydactylia, with which myositis ossificans progressiva is preëminently associated, and for claiming for this deformity an endogenetic cause.

We know that the "anlage" of the carpus consists of three parallel tissue rays (gewebstralen) which divide again in several parts of which the radial (or relatively tibial) and medial continue into the first two fingers (toes) and of the ulnar (fibular) ray which traces its origin secondarily from the ulna (fibula), issuing lateral branches from the fourth and fifth fingers, also for the basal elements of the third finger. Thus we see at first the "anlage" of first and second fingers, later of the third and last of the fourth and fifth. We find a certain relationship in the extent of deformities according to this normal mode of development.

The following references are only an approximately complete list of the literature of this deformity, and in order not to devote too much space to this section of my paper, the cases will be but briefly quoted:

M. Leboucq[74] mentions a case of bilateral symmetrical brachydactylia of all the fingers.

E. W. Roughton[75] demonstrated a case of brachydactylia of the right big toe in a 30-year-old man, and of the fourth toe on left foot. In the second case the brachydactylia affected the third, fourth and fifth toes of both feet.

Julius Sternberg[76] registered two cases; the first showed brachydactylia of both third fingers and both third toes; the second brachydactylia of both fifth toes.

E. E. Goldman[77] describes a case of brachydactylia of the third, fourth and fifth fingers of the right hand, and of the fourth and fifth fingers only on the left hand. The same patient showed symmetrical brachydactylia of both fourth and fifth toes.

L. Freund[78] reports a brachydactylia of both fourth fingers of a young man. The distal epiphyses were shortened and atrophied.

Rieder[79] mentions a man with an interesting case of combined brachydactylia of

[74] Brachydactylie et Hyperphalangie symétrique de l'Index et du Médicus Bulletin de l'académie Royale de Médecine de Belgique. Tome x, No. 5, 1896, p. 345. (This case had been presented already to the Med. Soc. of Ghent by Dr. M. Colson in 1883 and published in Pfitzner's essay on this subject.)

[75] Lancet, July 3, 1897.

[76] Symmetrische Verkuerzg. d. 3t. Mittelhdknoch. Wiener klin. Wschrft. 1902, No. 41, p. 1060.

[77] Beitrag. z. Lehr v. d. Missbldgn. d. Extrm. Beitraege z. klin. Chir., Bd. vii, 1891, p. 239.

[78] Die Brachydaktylie durch Metakarpalverkuerzung. Ztchrf. F. Heilkde. 1906, Abth. f. Chir. etc. Bd. xxvii, p. 129.

[79] Ueber gleichzeitiges Vorkommen von Brachy- und Hyperphalangie a. d. Hand. Deutsch. Arch. f. klin. Med., Bd. 66, 1899, p. 330.

the fourth finger of the right hand and hyperphalangia (three phalanges) of the thumb of the same hand. The daughter of this man had syndactylia of the right hand and hypophalangia of all other extremities.

Wagner[80] published the case of a woman who showed a bilateral brachydactylia of the third and a unilateral of the fourth fingers.

Hochheim[81] adds the case of a woman 18 years old with a bilateral brachydactylia of the third, fourth and fifth fingers and a brachydactylia of third and fourth toes, with shortened distal epiphysis and an imperfect development in breadth. A brother had a brachydactylia of the fourth right toe.

G. Joachimsthal[82] communicates two cases from his experience. He observed a woman of 27 years with a bilateral brachydactylia of the fourth and fifth and a brachyphalangia of the second and third fingers, and two women of 29 and 22 years, sisters of the former, with the same deformities, although not quite so pronounced. The only brother is normal, but has a daughter 1 year old with the same defects, and a son of the older sister had the identical deformities. In the same article were the very interesting cases of a young man with a bilateral brachyphalangia (middle phalanx) of the second and fifth fingers, whose mother was equally afflicted; a case of a girl 12 years old with a bilateral brachydactylia of the fifth finger, and of two women, 25 and 31 years old, each with bilateral brachydactylia of the fourth fingers.

From Cerechellis Institute, Carlo Fossetti[83] reports an observation of a bilateral brachydactylia of the fourth finger in a woman of 28 years. The distal epiphyses of the fourth metacarpals were shortened and atrophied.

Alfred Machol[84] describes the following cases from the Breslau clinic: Girl 17 years; brachydactylphalangia of the third finger of left hand. Besides a shortening of the third metacarpal bone, there existed also one of the basal phalanx. The distal epiphyses of the third metacarpal and the proximal one of the basal phalanx are rarefied and atrophied and demonstrate the cause of the deformity.

Woman of 34 years, shows a symmetrical brachydactylia of the fourth fingers and brachydactylia of the right fourth toe. The distal epiphysis of the fourth metacarpus is round like a ball and sits squarely on the diaphysis. The osseous structures of both metacarpal phalanges are rarefied.

A girl of 30 years with a unilateral brachydactylia of the fourth left toe. In this case also the osseous substance of the deformed toe is rarefied.

A girl of 12 years had scarlatina when two years old, followed by a paresis and debility of the entire right leg. Circumference over entire leg about 2 cm. less than the correspondingly right side. Brachydactylia of fourth toe of same side, osseous substance atrophied. Also smallness and atrophic state of phalangeal epiphysis, which may, however, be attributed to the generally impaired nutrition of that leg.

Boy 12 years old; poliomyelitis anterior when three years old. Pes varus sinister. Left foot slightly smaller than right. Brachydactylia of the fourth left toe. The osseous substance of left fourth metatarsus atrophic and bone-tissue rarefied. Distal epiphyses likewise compressed and squatty.

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[80] Beitrag zur Kenntniss der Brachydactylie. Fortschr a. d. Geb. d. Roentgensc. 1903, Bd. vii, pp. 94-98.

[81] Fall von Brachydaktylie. Fortschr. a. d. Geb. d. Roentgenstralen, 1904, Bd. vii, S. 273-76.

[82] Die angeborenen Verbildungen der oberen Extremitaeten. Ergaenzungsheft 2 d. Fortschritte auf dem Geb. d. Roentgenstralen. Hamburg, 1900.

[83] Brachidattilia simmetrica della mano. La clinica chirurgica 1914, No. 11, p. 239.

[84] Mitteilungen a. d. Grenzgebieten d. Medizin & Chir. (Mikulicz Gedenkband) Jena, 1907, p. 712.

Faulhaben[85] observed in a patient 22 years old symmetrical bilateral brachydactylia of the fourth and fifth fingers.

Fontana and Vachelli[86] encountered a most interesting triplicate of cases concerning two sisters, 16 and 12 years old, each with bilateral brachydactylia of the fourth fingers, and their mother, who showed this deformity on the left side only. Another woman, who did not belong to this family, also had a brachydactylia of the left fourth finger.

Lunghetti[87] relates the case of a man 32 years old with brachydactylia of both third fingers and brachyphalangia of the middle phalanx of the fifth fingers.

Kenyeres[88] saw a man of 37 years with symmetrical brachydactylia of the fourth fingers. The terminal phalanx of the left thumb was also about one-third shorter than that of the other side.

A. Mosenthal[89] describes (1) a brachydactylia of a young girl 20 years old affecting the fourth and fifth metacarpals of both hands (mother said to have had the same deformity), and showing a marked atrophy of these bones. (2) The cases of mother and daughter both with bilateral brachydactylia of the first metatarsus, with abduction of big toe (hallux varus).

Erich Ebsten[90] communicates in the same article an observation of brachydactylia of the left ring-finger and another case of a brachyphalangia of all four fingers of the left hand caused by the shortening of the middle phalanges only.

Similar cases of familiary heredity deformities of the fingers since the observation with the X-ray are but few, Julius Drey[91] had occasion to observe four generations of a family, the various members of which (thirteen normal, twelve defective) suffered from hypophalangia and digital synostosis.

Rich Pott[92] enjoyed a similar opportunity of observing in pre-X-ray times unmistakably hereditary defects in four successive generations. The members of this family, of which nine were normal, showed in five oblique nail phalanges of thumbs, three double end phalanges and three double entire thumbs (six complete fingers on each hand).

H. Drinkwater[93] reports two cases of hereditary transmission of digital deformities, members of both familes suffering from brachyphalangia. The deformity was caused in both families from an absence of the second phalangeal epiphysis, at the base only in one family, who showed in four generations twenty-six normal and twenty-one short-fingered members; the other in as many generations, with both epiphyses defective, had ten normal and nine short-fingered ones. This latter family was peculiarly interesting, as one of its normal married women had practiced illicit intercourse with a short-fingered man. She gave birth to one short-fingered son, who

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[85] Deutsche mediz. Wochenschr. 1904, p. 1454.

[86] Archiv. d. Ortoped. 1902.

[87] Sopre un caso brachydattilia simmetrica della mano. Archivio di Ortopedia Febr. 1902, p. 52.

[88] Fortschrite a. d. Geb. d. Roentgenstralen. Bd. ix, 1905-6, p. 351.

[89] Eleventh German Orthopædic Cong. Einige Faelle v. Brachydactylie. Verhdlgn. d. deutsch. Ges. f. orthop. Chir. 10th Congr. 1911, p. 36.

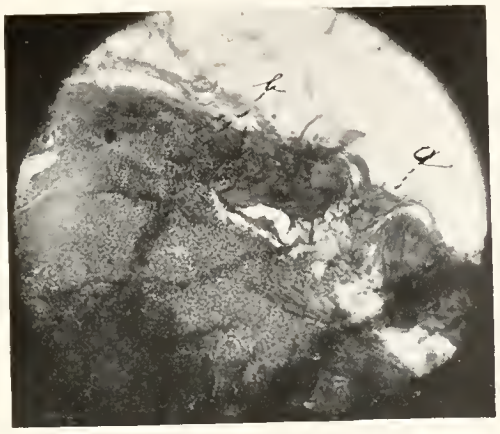
[90] Zur Aetiologie d. Brachydaktylie. Mitthelign. a. d. Grenzgebieten d. Mediz. und Chir. Bd. 21, 1910, p. 531.

[91] Hereditaere Brachydaktylie kombinirt mit Ankylose einzelner Fingergelenke. Festschrift gewidmet M. Kassowitz, p. 34, Berl., Julius Springen.

[92] Ein Beitrag. z. d. symmetr. Missbildgn. d. Finger & Zehen. Jhrbch. f. Kinderhlkde, Bd. xxi.

[93] Account of Family showing Minor Brachydactylia. Journal of Genetics, vol. ii, p. 21, 1912-13, p. 21.

A



B

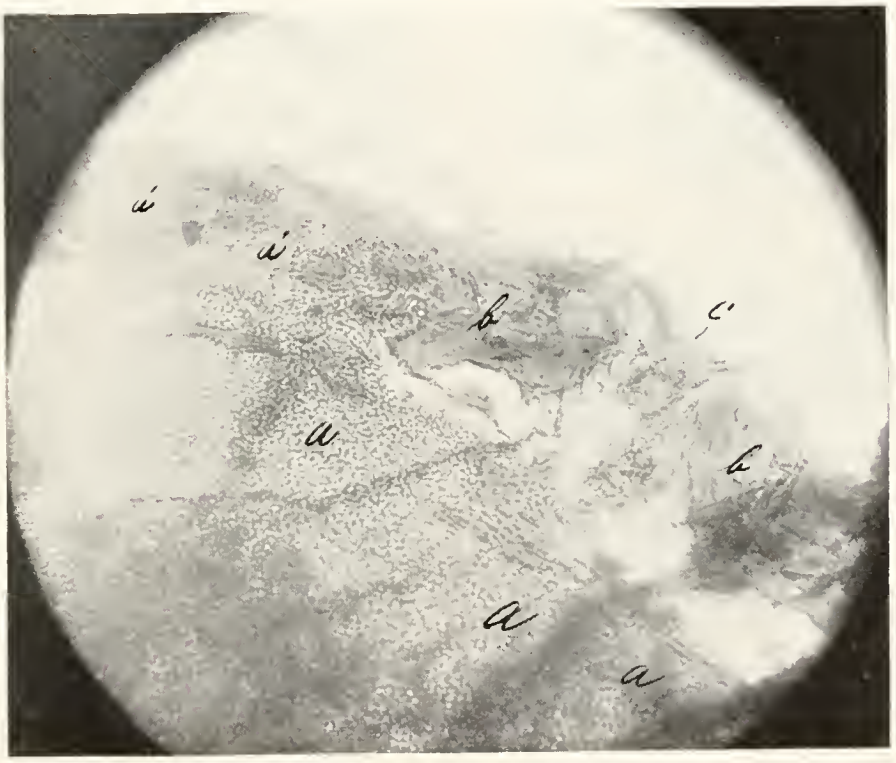


FIG. 35.—A, Connective-tissue bridge and predominant participation of erythrocyte in bone formation. *a*, connective-tissue bridge; *b*, erythrocytes participating preëminently on ring in bone formation (oc. 2, obj. 8). *B*. Connective-tissue bridge and predominant apparent erythrocyte participation in formation of ring (enlargement of Fig. *A*). *a*, erythrocytes; *a'*, erythrocytes in apparent participation of formation of ring; *b*, ring; *c*, connective-tissue bridge.

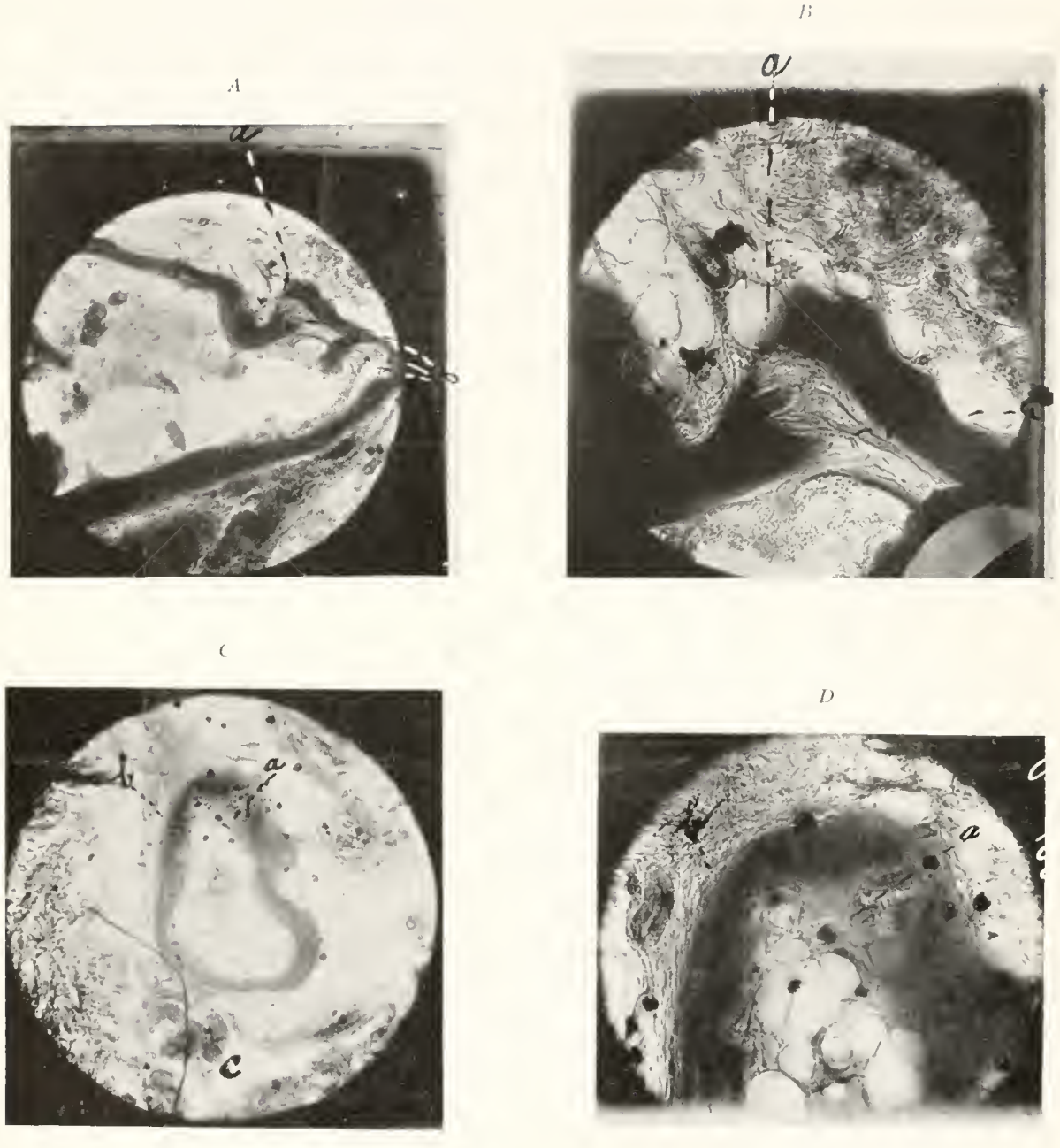


Fig. 36.

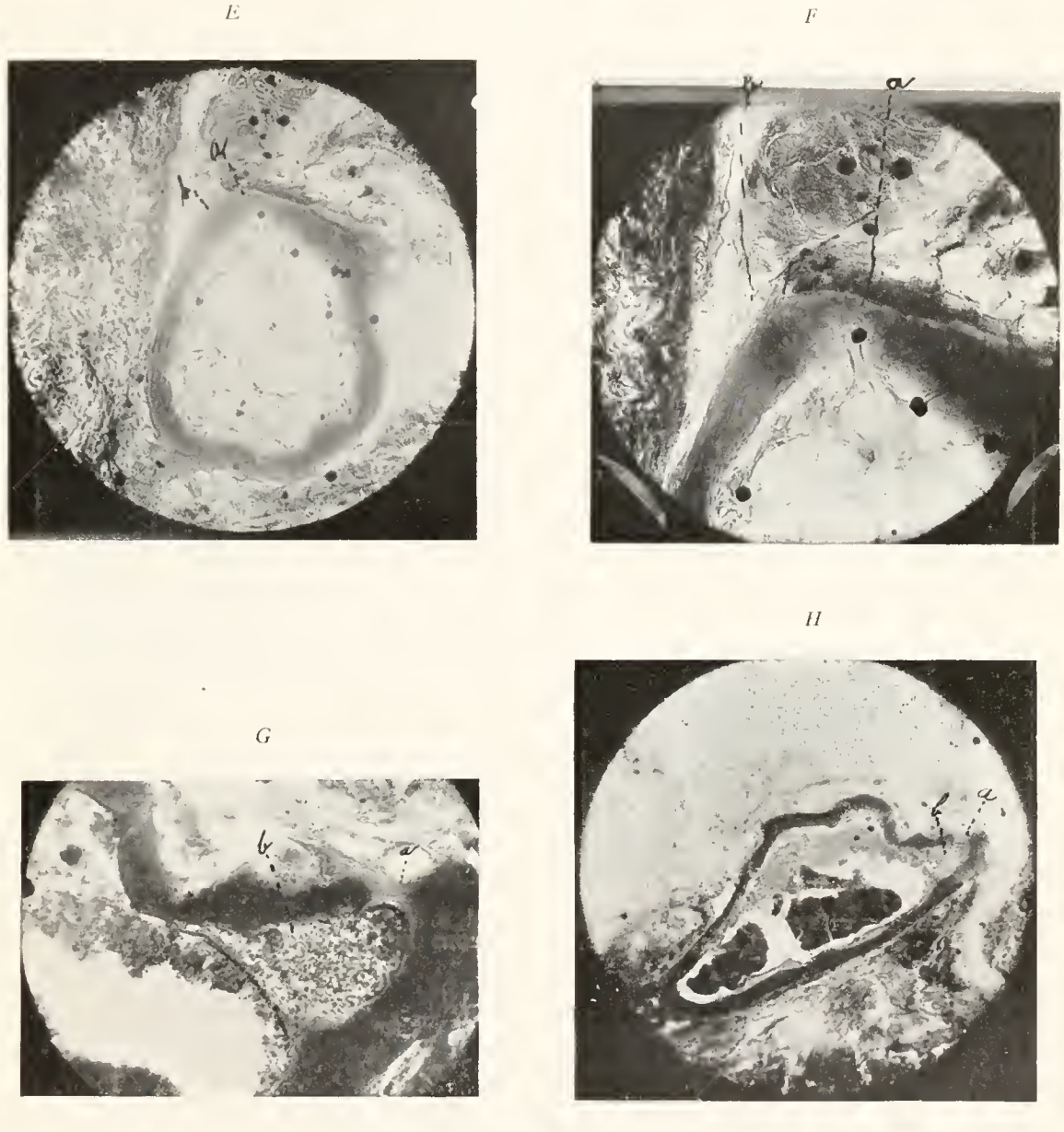


FIG. 36.—Connective-tissue bridge with projection inside. *A*, *a*, connective-tissue bridge; *b*, connective-tissue projection inside ring (oc. 2, obj. 16). *B*, *a*, connective-tissue bridge; *b*, connective-tissue projection inside ring (oc. 2, obj. 4). *C*, *a*, connective-tissue projection toward inside of ring and bridge; *b*, connective-tissue participation in bone formation; *c*, suderiferous gland (oc. 2, obj. 16). *D*, *a*, connective-tissue projection toward inside of ring and bridge; *b*, connective-tissue participation (oc. 2, obj. 4). *E*, *a*, connective-tissue bridge; *b*, connective tissue participation in bone formation outside of ring (oc. 2, obj. 16). *F*, *a*, connective-tissue bridge; *b*, connective-tissue participation in bone formation outside of ring (oc. 2, obj. 4). *G*, a, connective-tissue bridge: *b*, erythrocyte participation (oc. 2, obj. 16). *H*, *a*, connective-tissue bridge, *b*, erythrocyte participation (oc. 2, obj. 4).

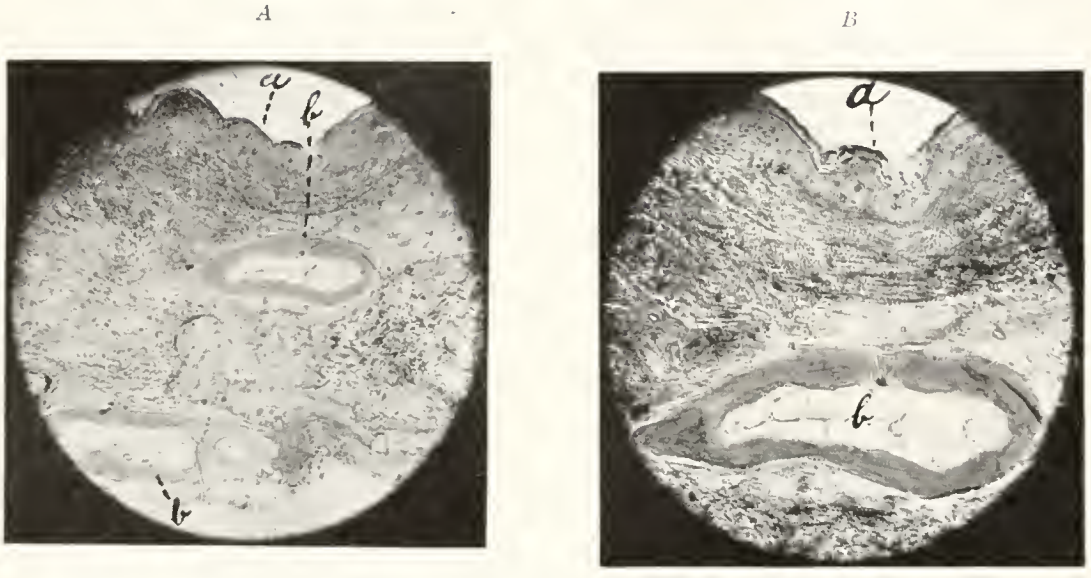
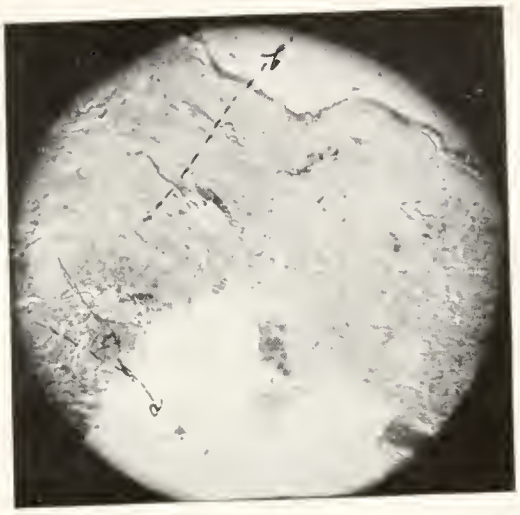
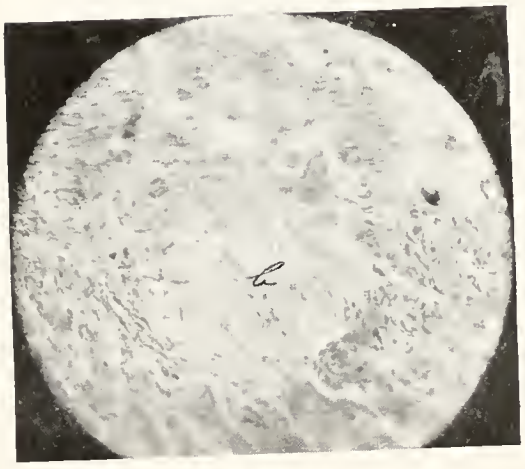


FIG. 37.—Bone rings with empty centre near epidermis. *A*, *a*, epidermis; *b*, emptied bone ring (oc. 2, obj. 2 *). *B*, *a*, epidermis; *b*, emptied bone ring (oc. 2, obj. 8).

A



B



C

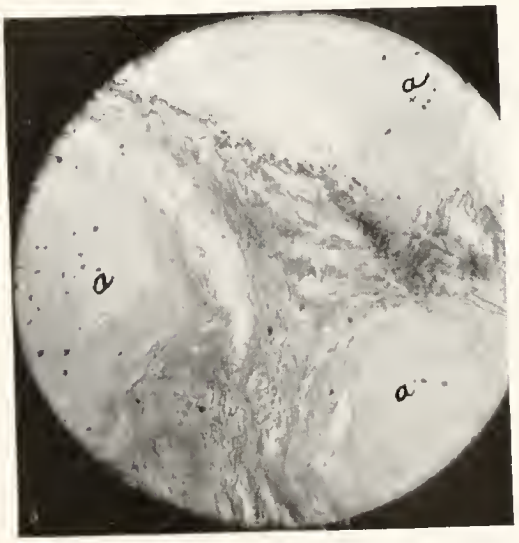


FIG. 38.—Initial hemorrhage at the side of three solidified bone formations, all close to epidermis. *A*, *a*, two of the bone formations; *b*, hemorrhage (oc. 2, obj. 2 *). *B*, *b*, hemorrhage (oc. 6, obj. 16). *C*, *a*, the three bone formations in part (oc. 6, obj. 16).

FIG. 30.—Right thoracic extremity of dog (anterior view). *a*, head of humerus in glenoida fossa; *b*, ossifications—compare diminutive first metacarpal and first phalanx of thumb with those of other digits.

FIG. 40.—Right thoracic extremity of dog (posterior view). *a*, head of humerus in glenoid fussa *b*, mass of ossification; *c*, location of thumb anteriorly.

FIG. 41.—Longitudinal section through right wrist and carpal bones.

in his turn was the ancestor of all the other short-fingered progeny, so that in this family the hereditary defect was limited to the side-issue of the hired man.

Philip W. Mathews[94] had the extremely rare opportunity to observe a hereditary true brachydactylia through five generations. All the afflicted members had shortening of the third, fourth and fifth fingers on both hands, with broadening of the third phalanx of both fourth fingers.

Mario Bertolotti[95] gives another very remarkable report of three generations of a brachydactylic family, with ten out of a total of fifteen members showing a brachydactylia of the third fingers; he enters into the developmental history of the metacarpal bones and refers to Meckel, who came to the conclusion that the first metacarpal, judging by its location and peculiar service, and especially owing to its mode of ossification (being the only one of the metacarpal bones which has its epiphysial nucleus like all the phalanges on the proximal side), should be considered the first phalanx of the thumb. Radiography shows that at times one encounters in boys from 5 to 15 years an accessory nucleus on the distal side besides the old classical proximal.

Ludwig Freund[96] published the first of such cases and showed that in about 24 to 30 per cent. an additional distal special epiphysiary nucleus to the usual proximal one in the first metacarpal (also the first metatarsal in myositis ossificans progressiva) may be seen. He also found an occasional extra proximal nucleus in the second and fifth (in our case in the fourth) and that the atrophic character of such nucleus formation shows a regressive anatomical variation.

According to Dubreuil-Chambardel[97] this ossifyng nucleus in a distal epiphysis of the first metacarpal was present in eighty-two cases out of a total of 240 X-ray examinations. It commenced to appear in the third year and the complete ossification was reached at about the thirteenth or fourteenth year.

Freund[98] has observed the accessory variation now and then on the proximal side of the second, and, even more rarely, of the fifth metacarpal bone. These men all believe that the choice of epiphysial nuclei of the metacarpal bones depends upon their separate physiological functions. Therefore the proximal site in the first metacarpal depends upon the adaptation of this metapodical process to the functions of the thumb. To support this hypothesis Bailleul[99] as well as Dubreuil-Chambardel have each published a radiograph of a case of a triphalangeal thumb, where the first metacarpal showed its nucleus of ossification only on the distal side.

According to Koehler[100], however, this anomaly represents a certain regressive type met with in the cetaceæ and other water-mammals, where the thumb and big toe have two epiphysial ossifying nuclei. He also quotes Henle as having demonstrated from comparative anatomy that the metacarpal bones had originally two nuclear epiphyses at the same stage of development, a proximal and distal one, although with the human the rule is the distal one for the four last and the proximal one for the first metacarpal. Cruveilhier and Gegenbaur hold the same opinion. They draw attention to the curious anatomical conformation of certain

[94] A Case of Hereditary Brachydactylia. Brit. Med. Journ., Oct. 31, 1908, p. 969.

[95] Contributi radiolog. allo studio della sciluppo osteogenetico dei metacarpei. La Radiologia medica. Marzo, 1915, vol. ii, Fasc. 3, p. 105.

[96] Ueber Pseudoepiphysen. Ztschrft. f. Morphologie & Anthropologie, vol. viii, 1905, p. 87.

[97] Sur le development du premier rayon digital; Gazette medicale du Centre, p. 288, 1913.

[98] Op. cit.

[99] Ossification du Ir metacarpein. Societe de Biologie. March, 1911.

[100] Vollzaehlige proximale metakarpale Epiphysen. Fortschr. a. d. Geb. der Roentgenst, vol. xix, p. 279, 1912-13.

cetaceæ, balaenidæ and sureniæ, who present double epiphyses of the four last metacarpal and an evident trace of an additional distal epiphysis in the first.

Freund[101] confirmed these statements by his own investigations of various cetaceæ. A double epipyhsial nucleus in human metacarpals appears, therefore, in the light of a reversive type or mongolism.

Koehler[102], who advocates this view, supports it with the report of a unique case in medical literature. A patient 12 years old, suffering from infantile myxœdema, showed, in the X-ray picture of his hand, a development of his carpal bones corresponding to that of a child of three to four years, and all the epiphysial nuclei of his five metacarpal bones were on the proximal side.

Bertolotti supplements this with a radiographic discovery in the course of examination of his above-mentioned family. He found the brachydactylic members showing pseudo-epiphysial basal nuclei of their second metacarpal bones.

We cannot close this discussion of functional adaptation of nuclei of ossification with regard to the first metacarpal bone without adding here an observation pointing apparently to the opposite direction. W. Rieder[103] published the case of a family of eight persons in its third generation, showing bilateral hyperphalangia of thumbs (first metacarpal and three phalanges). They were father, 45 years old; daughter, 19 years old (first marriage), and her two daughters, 12 and 11 years; also her son, 4 years old, all three from her second marriage.

This array of facts refutes, we believe, conclusively any attempt to explain the presence of brachydactylia from exogenic causes, either by amniotic bands or intra-uterine pressure. The overwhelming evidence of the hitherto unused material of myositis ossificans progressiva, showing in practically every true case the accompanying feature of symmetrical brachydactylia of fingers, toes or both, does not seem necessary to prove that defects in the primary " anlage " must be held responsible for this deformity; a view which Burgerhout[104] expresses very strongly in his dissertation.

We cannot enter here into the more subtle questions of mongolism or atavism, which present many tempting reasons for discussion, but stand firmly for the adoption of primary endogenic developmental causes for brachydactylia. To this extent we may expect now to clear the way for a possible later answer to the ever-open question for final causes. Even to the location for the embryonic layer which had been charged with the formative fault—our case furnishes a new aspect. Rolleston[105] as well as Morley, Fletcher[106] and others are united in singling out the mesoblast as the culprit, Rolleston calling it the skeletal layer with its subdivision of the muscular. My case shows bone-formation also in the skin and draws for the first time this outer layer into participation of the pathological process. As our present

[101] Op. cit.

[102] Op. cit.

[103] Eine Familie mit dreigliedrigen Daumen. Zeitschr. f. Morphologie und Anthropologie. 1900, p. 177.

[104] Op. cit.

[105] Op. cit.

[106] Op. cit.

views do not sanction the strict division of embryological layers as the exclusive formative agents of distinct types of tissue, it may perhaps be surmised that the pathogenetic cause exerts its influence at a very early period of embryonic life, before any distinct differentiation has begun.

We believe a similar origin may be claimed for the osteogenetic hemorrhages. In my series of microscopical pictures a gradual progression is shown from the primary hemorrhage into the corium to the osseous growth by either the fibrocellular or hyaline cartilaginous or the osteoblastic route to the final development into bone tissue. So clear is this progress that in some places the pictures appear to indicate a direct metamorphosis of the erythrocytes into the fusiform osteogenetic cells (fibroblasts). The entire course of events from primary hemorrhage to finished bone lies illustrated before our eyes, but we want to know the reason for these small hemorrhages and here we have to recur to the endogenetic fault in the "anlage" and have it extended to the capillary and minor peripheral blood-vessels.

The same developmental disturbances may, on account of the identity or close proximity of the basic tissue for both, easily affect the vascular together with the skeletal systems. The fingers or toes present, as endings of the skeletal system, anatomically similar relations, as the smallest blood-vessels and capillaries, as the last endings of the vascular system, in the integumental parts. A defective wall-construction of these two may account for a considerably lessened resistance in the integumental tissue, skin and muscular, to slightest traumatism, or variation of blood-pressure, and a consequential easy rupture with resultant circumscribed hemorrhages. Thus an acceptance of the theory of a simultaneous "anlage-defect" in the vascular as well as in the skeletal system may give us a better understanding for the causes underlying the early stages of the bone-forming process in this disease, which our microscopic sections so fully illustrate. We shall not go any farther in our explanatory attempts and refuse to go "a'sailing upon unknown seas" with Maunz[107], who, endeavoring to explain his views of formation of bone, asserts that with the anlage of the muscular system and the osseous skeleton, embryonic osteoblastic tissue-germs have been wandering about and deposited in such places of the muscular connective tissue, tendons and fasciæ, where later the osseous formations start. Nor shall we enter into a discussion with the other theorists, such as Goeschen[108], who, after referring to the peculiar calcium metabolism (?) of the myositis ossificans patients as stimulating the ossifying process, finally adds: "Up to date the acceptance of a disposition to such ossification cannot be entirely dispensed with." While we have offered a tentative explanation of the pathological anatomical development in line with the microscopical pictures, we must admit our complete ignorance of its more remote causes.

The therapeutic part of our theme is very brief. In the early period

[107] Op. cit.

[108] Ueber Histologie & Pathogenese der circumscripten Muskelverknoechrg. (Jena 1913. Gustav Fischer.)

of its history, the disease was attacked with all sorts of remedies, alteratives, solvents for calcium supposedly retained in the system by faulty metabolism, the inevitable iodide of potassium, etc.; every one of them with more or less marked success observed solely by its original author but pronounced a complete failure by every other follower. The favorable result of thiosinamine injections reported by Boseck[109] has never been achieved by anyone else; moreover, his alleged case of myositis ossificans progressiva lacks every convincing feature of deserving such classification. In myositis ossificans traumatica Aizner[110], Nicolai[111] and Grosskurth[112] have reported favorable action of the same drug (fibrolysin). This disease, however, disappeared often spontaneously, so the therapeutic effect should not be unreservedly endorsed. Lately Nové-Josserand and René Horand[113] have earnestly recommended an X-ray treatment for the progressive form. They report excellent results in their case by a series of twenty-six treatments, all below five Holzknecht. I had the abdomen of my case treated in this way without the least effect, except perhaps to stimulate new growths.

In summing up, the conclusions of this treatise are as follows:

CONCLUSIONS

1. Myositis ossificans progressiva is a misnomer for a pathological process of atopic ossification starting primarily in the connective tissue of the exoskeleton and involves in its further progress the tissues of the endoskeleton. It would better be named fibrocellulitis ossificans progressiva. The disease nearly always commences at an early period of life and shows as additional characteristic symptoms a microdactylia of fingers and toes, and in some cases a more or less pronounced degree of infantilism.

2. Similar to the primary hemorrhage of the traumatic myositis ossificans, the introductory feature of the pathological process in the progressive form is an initial intercellular capillary hemorrhage which starts the growth of newly-formed connective tissue surrounding the hemorrhagic extravasation without any previous hypertrophy of the adjacent connective tissue.

3. Calcification and ossification take place at first at the newly-formed connective-tissue strands encircling the hemorrhagic mass, and with few exceptions from within outward.

4. There is an apparent active participation of the enclosed erythrocytes in the bone-formation; they change their round form to a spindle-cell shape and seem to become nucleated as they approach the osteogenetic edge of the encircling ring and merge into its substance.

5. The microdactylia has an endogenetic origin and is the result of an "anlage" defect of the carpal and tarsal tissue rays ("gewebsstralen"),

[109] Zur Heilg. der Myositis ossificans progressiva geheilt durch Fibrolysin. Muench. mediz. Wochschr., 1906. p. 2350.

[110] Muench. mediz. Wochschr., vol. lvi, part 1, p. 756.

[111] Deutsche militaeraerztl. Zeitschr., Heft 18, 1907.

[112] Deutsche militaeraerztl. Zeitschr., Heft 18, 1908.

[113] Op. cit.

causing thereby an imperfect development of the last endings of the extremities in one part or other.

6. The initial hemorrhage is probably due to a similar cause affecting the integrity of the last endings of the vascular system, the capillaries, from where the hemorrhage starts.

7. The calcification and consequent ossification of the newly-formed connective tissue is due directly and indirectly to the primary hemorrhage. The hemorrhagic effusion with its calcium content starts the first deposit of lime and indirectly stimulates its continuation by a nutritive impairment of the surrounding tissue from pressure of the extravasation.

8. Investigations made under scientifically correct premises show there is no reason for believing in a diminished calcium excretion or a calcium retention in this disease.

9. No therapeutic measures against the products or the progress of the disease have proven of any value.

An agreeable duty has yet to be discharged. My heartiest thanks are due Miss Ophuls and Mrs. Hurwitz, the librarians of the Lane and U. C. Libraries, for their untiring help in providing the literature from their own and the Surgeon General's library; to Doctor Israel, from the latter institution, for the translation of the Slavic papers; and to Mr. Ginsburg, the roentgenologist of the Mt. Zion Hospital, for his patient and indefatigable photographic work.

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